Haemophilia of the third age

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Over the years there have been many attempts to define the different ages of man. One can think of the different problems associated with haemophilia and come up with the problems associated with the different ages of a person with haemophilia (PWH). The first age is childhood, the second adulthood, the third age is the period after retirement whilst for some individuals there is a fourth age of partial or full dependency. In childhood the problems of haemophilia include the diagnosis, introduction of treatment, prophylaxis in severe and some moderate individuals, dealing with inhibitors in previously untreated patients, compliance with prophylaxis and schooling. In adulthood the problems will depend on whether the PWH grew up with prophylaxis where almost normal health is possible or whether they grew up in the on demand era when joint disease is invariable with, in many persons, HIV and/or HCV infection.

This editorial was conceived when on a single day we dealt through our haemophilia centre with inherited bleeding disorder patients and the following problems: dementia, confusion, falls, visual impairment, use of antiplatelet drugs following coronary artery stenting, atrial fibrillation, stroke, hypertension, renal impairment, advanced arthropathy as well as acquired haemophilia in an individual with congenital haemophilia. As haemophilia professionals we now infrequently have to advise or treat severe PWH with concentrates outside the setting of surgery. In practice, all our severe and many moderate PWH have their concentrates delivered to their home by a third party, are able to self infuse, and so rarely trouble the haemophilia centre for advice on how to treat new bleeds. The result is that much of our time is now devoted to managing comorbidities associated with chronic joint damage, chronic infections and the problems of older age.

As individuals get older the risk factors for cardiovascular disease such as hypertension, obesity and diabetes mellitus become more prevalent(1). These factors are at least as frequent in PWH as in the general population but despite these, the risk of death from a myocardial infarction or stroke is less. Since the rate of atherosclerosis is the same, it is believed that the reduced cardiovascular death rate is due to smaller thrombi forming over ruptured atherosclerotic plaques(2). Two additional cardiovascular problems seen at increasing frequency are atrial fibrillation and acute coronary syndrome / angina where the problem is less about the acute management than the long term use of anticoagulant and antiplatelet medications (3).

Orthopaedic problems are a frequent feature of many adult PWH but two additional problems are encountered with increasing frequency in this age group, osteoarthritis and redo joint replacements. Osteoarthritis can affect almost any joint and replacement of these joints is more common. In practice there is no real difference in terms of the haemophilia management of a PWH having a joint replaced for haemophilic or osteoarthitic arthropathy. When joints initially started being replaced in PWH it was felt that patients may not survive long enough to need them to be redone. It is now fairly common to have patients requiring replaced joints to be redone with no obvious upper age limit. We have a 96 year old severe haemophilia A

patient attending our centre in whom we performed redo knee joint replacement surgery at the age of 92 years; he had that joint first replaced 20 years earlier.

In addition to the physical disability associated with haemophilic arthropathy, chronic pain and reduced muscle bulk may lead to increasingly abnormal gait which in turn leads to an increased risk of falls and secondary injury (4). The increased risk of reduced bone mineral density in PWH also raises the chance of fractures occuring secondary to falls.

The ageing PWH can experience difficulties in gaining venous access through numerous factors. Multiple venepuncture attempts may heighten patient anxiety and suffering, delay vital treatment and increase costs. PWH may switch back to on demand regimes in preference over prophylaxis in the hope of reducing the requirement to access veins. Small, fragile or hidden veins can lead to difficulties, and collapsed veins due to dehydration are especially problematic. In addition, reduced visual acuity and depth perception, reduced manual dexterity and coordination may all contribute to reduced independence with clotting factor concentrate administration.

The PWH who needs to retire early through ill health may feel the loss of a sense of purpose and wellbeing that an individual gains from everyday contact with work colleagues. The reduction in the wider social network can lead to a refocus on family relationships. In adulthood, the focus of the family network is an individual's partner, and ultimately their children. The effect of the loss of spouse, family and friends through bereavement or ill health has a specific and significant impact on an individual's personal network. Psychological problems such as dementia and depression require the support and dedication of a close family network. Bereaved individuals, childless people, unmarried and those who live alone may be at particular need of additional support.

As age advances and patients enter the fourth age of dependency some specific problems can arise as some residential homes refuse to accept individuals who may require intravenous concentrate infusions or who are chronically infected with HIV or hepatitis C. It is sometimes necessary to commence prophylaxis as individuals depend on nursing home personnel that are unable to give on demand intravenous injections.

In this article we may have given the impression that there is clear separation between the different ages but this is often not the case. The end of the first age was traditionally the end of the teenage years but in many Western countries it is now increasingly the age of going to University, leaving home or gaining first full employment. Although we suggested that the third age starts on retirement this happens at different ages in different countries and within a country varies between individuals and over time. The idea is that this occurs around the 60-65 year mark as older age comorbidities begin or start to increase at this age. Haemophilia care is no longer about treating bleeds but rather about treating the individual as a whole with all their accumulating medical comorbidities.

Declarations of Interest
The authors declare that they have no relevant conflicts of interest.

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Table 1: Some of the issues at the different ages of a person with haemophilia

First age: Childhood
Diagnosis
Introduction of treatment
Prophylaxis
Inhibitors
Adherence to treatment
Schooling and sport

Second age: Adulthood
Arthropathy
HIV and HCV coinfection
Prophylaxis
Inhibitors in some patients

Third age: Retirement
Cardiovascular disease
Renal disease
Hypertension, obesity, diabetes
Increasing tendency to falls
Decreasing visual acuity
Dementia
Malignancy
HIV and HCV coinfections
Mobility
Arthropathy
Bone health
Loss of social network

Fourth age: Dependency
As in third age but worse
Seeking care/support outside the home or hospital