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Review Article

Relationship between haemophilia and social status

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ABSTRACT

The impact of haemophilia and its treatment on social status has not been well studied, although research into the quality of life of patients with haemophilia has shed some light on aspects of social and role functioning. Studies conducted before the advent of safe and effective coagulation factor replacement therapy suggest that the haemophilia population was predominantly of low socioeconomic status with many social disadvantages, including high rates of disability and unemployment and low rates of marriage. Since the availability of purified factor VIII concentrates that could be used in a home-care setting and as prophylaxis, most research suggests that social status and well-being amongst children, adolescents, and adults with haemophilia is not compromised, and is comparable to that of the general population. Children and adolescents with haemophilia do not generally feel disadvantaged, although haemophilia-related issues at school and amongst peer groups do arise. Recent studies in adults show higher than average rates of marriage and cohabitation and the attainment of a generally good educational status, but, as in the past, employment rates remain comparatively lower. Social status amongst the elderly with haemophilia who may have developed severe disability as a result of their condition is poorly defined and has never been formally studied. Additional research is recommended.

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Introduction

The haemophilias are a group of life-long disorders associated with a chronic burden of morbidity that is worsened by episodes of acute bleeds and their consequences [1]. Before modern substitution treatment was introduced at the end of the 1960s, haemophilia often resulted in severe disability at a young age and premature death. The possibility of someone with haemophilia enjoying a full social life was non-existent or limited [2]. The availability of clotting factor concentrates and the introduction of prophylaxis and home treatment greatly improved quality of life for people with haemophilia and their families [2], and now, many individuals with haemophilia, especially younger patients, do not consider their condition to be a major burden [3].

The management of haemophilia needs to consider not only the patient's medical requirements but also their emotional and social needs, including those relating to social status – an aspect that is often overlooked in haemophilia studies. In its simplest form, "social status" refers to the position an individual occupies in the social structure, such as being a teacher or office worker. Social status is often considered in combination with "social role", which refers to the role an individual plays within their social group and in society as a whole.

Abbreviations: AIDS, acquired immunodeficiency syndrome; HIV, human immunodeficiency virus; SF-36, Short Form 36.

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Several factors help determine an individual's social status. In adults, income, education, and occupation have traditionally been considered core components of social status, with studies focusing primarily on objective measures, including social class identification [4]. In children and adolescents, social status is determined more by subjective perceptions of the individual's placement within the social hierarchy in terms of peer respect, academic or sporting success, and social "standing" [4]. In the elderly, social status is more likely to be determined by an individual's mobility, residential situation, and social support network; however, this has not been well defined in the literature.

Several different approaches to evaluating "social status" in haemophilia have been taken over the years. Quality-of-life studies, which utilize both generic and disease-specific questionnaires, have assessed certain components of social and role functioning, but do not specifically evaluate social status *per se* [5]. The Short Form 36 (SF-36), for example, which is one of the most commonly used generic instruments for assessing quality of life, quantifies physical and social functioning, and the ability of individuals to fulfil their physical and emotional "roles" [6]. Studies assessing social status in adults have evaluated marital, educational, and occupational status [3]; those in children and adolescents have focused on academic achievement [7] and perceived placement within the school community [4].

Impact of Haemophilia on Childhood Social Status

School attendance and academic performance can be adversely affected by haemophilia and its treatment [7], although studies in adults suggest that most individuals with haemophilia ultimately attain a normal, if not elevated, educational status [3].

Children with haemophilia under prophylactic therapy can benefit significantly from participation in physical and sporting activities, both in terms of their health and quality of life [8–11]. Prior to the introduction of prophylactic factor replacement therapy, children were discouraged from physical activity due to the risk of bleeds, resulting in some children having lower levels of fitness and strength than their peers [12]. Although, today, additional protection may be required when engaging in sporting activities, the benefits to children far outweigh the risks, including normalizing strength and fitness [11], enhancing self-esteem, increasing social contact, and encouraging a more "normal" way of life [10].

Interest in quality-of-life research in children with haemophilia is growing [13]. One of the most comprehensive assessments in children evaluated academic performance and certain aspects of quality of life, and found that the number of bleeding episodes was positively correlated with school absenteeism, which, in turn, correlated with lower achievement scores in mathematics, reading, and total achievement [7]. More recently, Gringeri et al. evaluated the health status and health-related quality of life of 318 children, aged 4–16 years, from six European countries and found that, although quality of life was broadly satisfactory, the younger children (aged 4–7 years) were negatively affected in the areas of "family" and "treatment", whereas older children had impairments in the social areas of "perceived support" and "friends" [14].

Impact of Haemophilia on Social Status in Adolescents

Adolescence is a critical time in a child's life in which he or she transitions between the social status of childhood, determined primarily by family status, and adult social status, which is largely self-determined [4]. Chronic illness during adolescence can have a profound effect on an individual's long-term health and well-being. Several studies are suggesting that chronic conditions and the demands of treatment regimens during adolescence can adversely affect growth and development, identity, mental health, relationships with peers, and engagement with education and employment [15]. In a study from Iran, 49% of 100 haemophilia patients aged 16–67 (mean age 28±9) reported that haemophilia had a negative impact on education [16]. Severity of disease and number of affected joints led to a negative attitude towards continuing education.

Despite this period of heightened social changes, the effects of haemophilia on social status in adolescents remains poorly studied. Most teenagers with haemophilia participating in a Scandinavian study conducted between 2003 and 2004 reported that they had the same opportunities as their peers in terms of engaging in normal teenage activities (such as playing sport, having a girlfriend, having sex, drinking alcohol, and partying); just over half (52%) had contact with other haemophilia patients, and most (94%) had told their friends about their condition, even though this was considered to be one of the most difficult aspects of having haemophilia, along with the demands of treatment and restrictions in physical activities [17].

Adolescence is also a period fraught with sexuality issues, and having haemophilia can intensify most of the normally encountered concerns [18]. Dating and sexual intimacy can be daunting prospects for teenagers with haemophilia, and issues associated with sexual desirability, sexual performance, and transmission of sexual diseases and genetics are common [18]. Many young men struggle with how to disclose their diagnosis to a potential partner, and they often need assistance with handling disclosure and with confronting their own doubts about becoming intimately involved with another person [18].

The choice of education and careers, which are considered key components of social status, becomes especially important during adolescence and early adulthood because of the need to avoid or minimize the possibility of bleeding. Individuals with haemophilia are generally advised to avoid careers that require strenuous physical labour or work in industries where injuries can occur. Bleeding episodes and related absences may put employment at risk if employers favour a "healthy" employee over someone with a "disability" or with a higher possibility of frequent absences from work. Thus, it is unsurprising, that some adolescents with haemophilia believe they have fewer work and career opportunities than others [17].

Young adults with haemophilia who acquired human immunodeficiency virus (HIV) have additional psychosocial and status challenges to contend with, often confronting social stigma and having to cope with increased financial burden, disruption of normal activities, and poor social relations [19,20]. The combination of haemophilia and HIV can be especially difficult in adolescence, with parents reporting high levels of pessimism about their children's futures, low levels of family integration, limited family opportunities [19], and intimacy concerns.

Such individual vulnerabilities may be further compounded by the fact that adolescence is often the time patients transition from paediatric to adult haemophilia care services; a process that is known to increase the risk of deteriorating health and quality of life in other chronic conditions [21]. Thus, a close cooperation between child and adult comprehensive care centres is paramount.

Impact of Haemophilia on Social Status in Adulthood

Adults with haemophilia contend with a range of complex psychological and social issues as a result of their condition. Despite the availability of factor replacement therapy, most adults still experience occasional bleeds, and those who did not have access to effective treatment early in life may still suffer disability in adulthood as a result of joint dysfunction and chronic pain [3,16,22,23].

There are a significant number of adults with haemophilia living with comorbid hepatitis and/or HIV/acquired immunodeficiency syndrome (AIDS), and these individuals have a higher burden of morbidity than other haemophilia patients in terms of impaired mobility, poor ambulation, and pain [1]. Studies suggest that individuals with both haemophilia and HIV/AIDS experience significant interference with work, education, and community responsibilities, and frequently