Diversity in Health and Social Care 2008;5:237–9

Guest editorial

Sickle cell disease: quality of care needs to improve within the NHS

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Next year witnesses the 30th anniversary of two significant developments concerning the delivery of care to those affected by sickle cell disease (SCD). The first ever NHS Sickle Cell Counselling Centre (in Brent, northwest London) was established in 1979, as was the Sickle Cell Society, a national voluntary organisation. These anniversaries provide a useful opportunity to reflect on the current provision of health services for this inherited blood disorder that affects an estimated 12 500 to 15 000 people, mainly from black and minority ethnic (BME) communities in the UK (Hobson, 2008). This compares with an estimated 8000 affected by the genetic disorder cystic fibrosis (www.cftrust.org), that primarily affects white individuals.

Abuateya and colleagues (2008) have recently produced an extremely informative review of the literature surrounding the education experiences of young people with SCD. They argue that the majority of studies on SCD have a clinical focus, and that debates on this and other chronic conditions rarely engage with the broader social context. While totally in agreement with these sentiments, the focus here will be on three recent developments that have had a particular impact on the quality of healthcare for those at risk of having children with the disorder and those who are living with the condition.

Screening for sickle cell disease

Sickle cell disease is a chronic life-threatening illness, and the complications, such as recurrent painful episodes, infections, acute chest syndrome and strokes in young children, are variable and unpredictable (de Montalembert, 2008). Embedding comprehensive services for SCD within national NHS strategy has been a major campaigning issue on all fronts and for many decades (Anionwu and Atkin, 2001). Early diagnosis of SCD to avoid preventable deaths among young children, from recognised complications such as pneumococcal infections, is one such example. Back in 1981 the Sickle Cell Society recommended 'that there be a national policy to screen newborn babies for sickle cell disease' (Sickle Cell Society, 1981). Twenty years later *The NHS Plan* (Department of Health, 2000) took a radical step in the provision of funds for a programme in England that included the screening of all newborn babies for SCD, regardless of ethnic origin. Complete rollout was accomplished in 2006, and about 300 babies are diagnosed with the illness each year (Streetly, 2008a).

While SCD is a condition that mainly affects BME groups, routine screening has identified higher than anticipated numbers of traits in white babies. Results of the newborn screening programme show that in England sickle cell disorders are as common as cystic fibrosis (Streetly *et al*, 2008). In addition, a more systematic approach to the offer of screening of pregnant women and their partners is to be completely rolled out in all parts of England before the end of 2008 (Streetly 2008b).

Improving the quality of care for those affected by sickle cell disease

Equally important is that all those found to be affected by SCD are assured of decent care, regardless of which part of the country they happen to reside in. Of concern is that progress has been considerably slower in this area compared with that of screening. The goal of comprehensive specialist centres, similar to those for haemophilia and cystic fibrosis, has still not been achieved. While a few NHS commissioners are supportive, a significant number of others still require convincing. Their sleep would be disturbed if they took the trouble to read the 2008 report *A Sickle Crisis?* from the National Confidential Enquiry into Patient Outcome and Death (NCEPOD) (Lucas *et al*, 2008), the findings of which are extremely unsettling.

The Enquiry focused on 81 patients with the inherited blood disorders of either SCD or thalassaemia who

had died between January 2005 and December 2006. Responses to requests for the case notes yielded a total of 55, each of which was then reviewed; the majority had SCD.

Many patients with SCD had been admitted due to acute pain, and required treatment with opiates which may depress respiration. Of the 19 patients who were being treated for severe painful episodes, nine had received excessive doses of opiates; five died as a consequence. Other known complications of SCD include renal failure. Lack of knowledge amongst nurses and doctors was demonstrated in the omission of standard observations such as respiration rate, fluid balance and the analysis of urine. Case histories describe how the neglect of such monitoring resulted in a failure to recognise the deteriorating state of health of some patients, with fatal consequences.

Standards of care for adults with SCD

The Sickle Cell Society recognised, as far back as 1981, the urgent need for education among health and other professionals involved in the care of patients with SCD. This was included in the 24 wide-ranging recommendations of a report entitled *Sickle Cell Disease: The Need for Improved Services* (Sickle Cell Society, 1981). As a founder member I can still recall the numerous appeals from family members, and sometimes health professionals, for the organisation to act as an advocate for patients whom they felt were receiving inadequate treatment. Occasionally the calls came following a death, and reading the NCEPOD publication resulted in those sad circumstances flooding back.

Surprisingly, until this year, there have been no treatment standards for adults with the condition. The Sickle Cell Society took up the cudgel three years ago, but members were shocked at their failure to obtain funds via the NHS. With monies obtained from a pharmaceutical company, in the form of an unrestricted educational grant, they brought together users and specialists to redress the situation. The *Standards for the Clinical Care for Adults with Sickle Cell Disease in the UK* (Sickle Cell Society, 2008) were launched at the House of Commons in July 2008 and can be downloaded from www.sicklecellsociety.org. It will be seen that they are not merely confined to healthcare and commissioning (e.g. specialist comprehensive centres), but also cover, for example, access to services covering housing, benefits and psychosocial advice.

The question, though, is how can such an important document be disseminated and implemented throughout the NHS so that the quality of care improves and deaths through negligence are avoided? A small BME organisation such as the Sickle Cell Society does not have the capacity to undertake this mammoth task alone and, indeed, should not be expected to do so. Last year it raised £300 000, in stark contrast to the Cystic Fibrosis Trust total of £11.5 million (Hobson, 2008). The NHS has a moral responsibility to redress historical oversights and oversee the implementation of the recommendations in both the NCEPOD report and the Standards of Care. In addition, funding is required for an educational programme for all relevant staff, in partnership with the Sickle Cell Society, the UK Forum for Haemoglobin Disorders, and the NHS Sickle Cell and Thalassaemia Screening Programme. It should include a learning resource for students planning to work within the NHS as nurses, doctors or other healthcare practitioners. This would require that the resource be routinely included in their pre-registration courses in higher education institutions, together with support for those tutors who know little about the subject. As a former professor of nursing I am aware, for example, that there are a significant number of nurses who have completed a three-year training course without ever having had a session on SCD. The regulatory bodies of healthcare education also have a responsibility to monitor whether the subject is embedded within the curriculum. A question or two in key assessments and examinations about care of patients with SCD could help to ensure implementation.

If steps of this kind were taken, there might be room for celebration of the aforementioned 30th anniversaries in 2009, a year that also marks the publication of the first NHS Constitution. After all, according to the consultative document launched in July 2008 (Department of Health, 2008), the first of seven principles that guide the NHS states that:

> The NHS provides a comprehensive service, available to all irrespective of gender, race, disability, age, religion or sexual orientation. It has a duty to each and every individual that it serves. At the same time, it has a wider social duty to promote equality through the services it provides and to pay particular attention to groups or sections of society where improvements in health and life expectancy are not keeping pace with the rest of the population.

Services for those affected by sickle cell disease come well within this duty, and should be on a par with those provided for individuals with cystic fibrosis.

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