Moving young people with sickle cell disease from paediatric to adult services

This article reviews data about transition from paediatric to adult services in patients with sickle cell disease, the most common inherited disease in the UK, and outlines how this has been addressed in a large UK sickle cell centre.

Sickle cell disease is an inherited blood disorder caused by a single base pair substitution of glutamic acid by valine at position 6 of the β-chain of haemoglobin, resulting in the abnormal production of haemoglobin S (HbS). The acute complications of sickle cell disease include recurrent vaso-occlusive painful crises and haemolytic anaemia, and chronic complications include stroke, chronic lung disease, pulmonary hypertension and renal failure. The term sickle cell disease refers both to homozygous sickle cell disease (HbSS) and the compound heterozygote condition where HbS is co-inherited with another abnormal haemoglobin; this includes HbS-β thalassaemia (co-inheritance with β thalassaemia) and HbSC (co-inheritance with haemoglobin C). The majority of patients in the UK with sickle cell disease now survive into adulthood, with over 98% surviving beyond their 18th birthday (Telfer et al, 2007). Cohort studies from the USA and Jamaica have reported median life expectancy to be in the mid-forties or mid-fifties in patients with HbSS, and as survival improves there is an increasing need for improved coordination of the transfer from paediatric to adult care (Platt et al, 1994; Wierenga et al, 2001).

Transition is defined as 'the purposeful, planned movement of adolescents and young adults with chronic physical and medical conditions from child-centred to adult orientated health care systems' (Blum et al, 1993). Poor transition services in patients with chronic diseases have been associated with unnecessary dependency of the young patients on their parents or guardians, developmental difficulties, psychosocial delay, lack of continuity of care, disengagement from services, and increased morbidity and mortality with subsequent additional health-care costs (Watson, 2000). Systematic reviews have shown few studies which specifically address these issues in the sickle cell disease population (Bryant and Walsh, 2009). This article reviews data referring to transition in all patients with chronic diseases and then discusses specific issues in adolescents with sickle cell disease and how this is being addressed by a single institution.

Dr Jo Howard *is Consultant Haematologist in the Department of Haematology, Guy's and St Thomas' NHS Foundation Trust, London SE1 9RT,* **Ms Tanya Woodhead** *is Medical Student, King's College London,* **Mr Luhanga Musumadi** *is Clinical Nurse Specialist in the Department of Haematology, Guy's and St Thomas NHS Foundation Trust,* **Dr Alice Martell** *is Clinical Psychologist in the Department of Paediatric Psychology and* **Dr Baba PD Inusa** *is Consultant in Paediatric Haemoglobinopathy in the Department of Paediatrics, Evelina Children's Hospital, Guy's and St Thomas NHS Foundation Trust, London*

Correspondence to: Dr J Howard

The psychosocial factors of adolescence

The demands of coping with chronic illness add to the challenges to adolescent development. Adolescents with sickle cell disease are more likely to show problems with adjustment (Lavigne and Faire-Routman, 1992) or internalize their thoughts and feelings leading to depression, suicidal ideation, anxiety, social withdrawal and low self-esteem (Williams et al, 2002). This may be caused by limitations on their prospects of future careers and achievements imposed by cognitive or physical disease complications. Sickle cell disease may also be accompanied by delayed puberty which can heighten self-consciousness and dissatisfaction with body image and may lead to ridicule by peers and consequent experience of social isolation at a time where peer relationships become increasingly important for psychological wellbeing. Adherence to medical regimens and regular attendance at outpatient appointments declines on entering adolescence (Ricker et al, 1998). Nevertheless adolescents with chronic illness who receive appropriate management and support are more advanced than other adolescents in their problem-solving ability.

The quality of interpersonal relationships with family members can have an important psychological and physical effect on the outcome of adolescents with chronic illness (Ricker et al, 1998). Parents have an important buffering effect on the potential stress caused by living with a chronic illness, but as the young people move towards adulthood this buffering effect may be lessened and the reality of living with sickle cell disease may become more apparent.

Adolescents who have successfully transferred to adult services describe feeling empowered, as if they have 'made a break' from their parents and their opinions are being listened to without the need for their parents to provide a safety net or buffer from bad news (Burlew et al, 2000).

Once adolescents have made the transition to adult services they are reported to be more content with the medical care they receive (Tuchman et al, 2008). They describe a difference in the way the two services are run and are happy with the efficacy of the adult services while missing the nurturing tendencies of the paediatric service.

Transition in patients with sickle cell disease

Several studies have used questionnaires or focus groups to identify the key challenges facing young adults with sickle cell disease during these transitional years (*Table 1*). As with other chronic diseases, young people with sickle cell disease have mixed feelings about transition, these include concern about leaving their paediatric provider who may have cared for them since birth, anxiety about moving to an adult service with new health providers who they have not previously met, and concerns about increased levels of self management and independence necessary in the adult service. Older adolescents may be looking forward to transition, and feel that they are ready to leave the paediatric service.

Families and carers also display concern about the young people leaving paediatric care and accessing adult care and about their ability to self manage (Telfair et al, 1994; Hauser and Dorn, 1999; Wojciechowski et al, 2002; Jones et al, 2007). Surveys in services without transition facilities have revealed high levels of awareness about the importance of transition, but equally high levels of anxiety about transition, and low levels of readiness for transition (McPherson et al, 2009). The lack of preparation for transition is an area for concern and there is a significant need for the development of transition services. Both adolescents and adults thought these should include information sharing and opportunity to meet adult health-care providers, in order to help with self management and educational provision. Practical issues about modalities of pain relief and how to access adult health-care providers were also raised (Telfair et al, 1994; McPherson et al, 2009). Concerns about funding of adult health care and the availability of adult providers are frequent concerns in American studies (Telfair et al, 1994). These concerns are less relevant in the UK, as the NHS funding system provides a more uniform delivery across the country.

A cross-sectional study of adolescents with sickle cell disease in the UK and the USA used validated questionnaires to assess their self-management skills. This showed no significant differences between the two countries, but did show that self efficacy (the extent to which individuals feel they have control over the events which affect their lives) was associated with age, educational level, psychological symptoms, knowledge about sickle cell disease and confidence with care, leading to the conclusion that transitional programmes in patients with sickle cell disease would be of benefit (Anie et al, 2005). Further studies looking at adolescents with sickle cell disease have shown that increased self efficacy is associated with fewer physical and psychological symptoms. Self efficacy was also linked to good self management, including drinking enough fluids (Clay and Telfair, 2007). Effective interventions to increase self efficacy during the transitional period will lead to smoother transfer to adult services and may lead to improved long-term self management, and these interventions should be included in transitional programmes.

A review in this area highlighted three key areas: lack of transitional support, limited transition programmes and transitional resistance to adult care. From this were drawn recommendations which include starting the transition process early, obtaining transitional support from the institution, the development of transition programmes with collaboration between the paediatric and adult provider and establishing relationships with patients who have already transitioned (Bryant and Walsh, 2009). The authors have drawn on many of these recommendations in developing their own programme.

There are very few transition programmes in either chronic disease or specifically in sickle cell disease in the literature. One example from the USA has described the use of a 'transition binder' which is given to children with sickle cell disease at the age of 13 years and contains skill checklists, tracking sheets for appointments, contact numbers, pain management plans and educational material. This department also offers a 'coaching through the emergency department' programme for 14–16-year-olds which helps adolescents to develop the skills necessary for communicating by themselves with health professionals, and enables them to take more responsibility for their health (Doulton, 2009). This programme has not been formally evaluated for effectiveness, and to the authors' knowledge, while there are several centres in the UK offering transition clinics, there are no comprehensive transition services for patients with sickle cell disease in the UK.

Addressing the needs of adolescents and their families: a single institution approach

It is estimated that around 80% of the 12 500 UK sickle cell population lives in London, with Lambeth being one

Table 1. Key issues for transition in sickle cell disease

Key issue	Management strategy
Issues with adjustment to chronic illness, leading to depression and low self esteem	Psychological support to help develop coping strategies
Concerns about moving from paediatric to adult provider	Introduction to adult team, tour of adult department, gradual move from paediatric to adult team. Transition team to provide continuity
Increased levels of self management and independence required in adult service	Health transition plan (transition passport) to assess readiness to move. Focus on increased self-efficacy. Education and information about sickle cell disease
Concerns about access to adult service and modalities of pain relief	Multiple meetings with transition team and adult team to address these issues
Need for flexibility in transition as cognitive impairment may decrease speed of transition	Start transition process early (12–14 years) and identify and act on individuals' transitional needs
Family anxieties leading to resistance to transition	Involve family in transition process including clinics, open days and departmental tours
Difficulties of first adult outpatient appointment and first inpatient admission	Transition team (clinical nurse specialist and psychologist) to provide support at these times

of the areas where it is most prevalent (Streetly et al, 2009). Guy's and St Thomas' Hospital, which includes the Evelina Children's Hospital, is situated in Lambeth and oversees the care of over 500 children with sickle cell disease. A transition clinic has been in place since 2001 (Okpala et al, 2002), but in response to national recommendations (Department of Health, 2003, 2006; NHS Sickle Cell and Thalassaemia Screening Programme, 2006; Sickle Cell Society, 2008) and following a survey of service users, the new strategy was developed. This included the appointment of a dedicated clinical nurse specialist and psychologist for transition and innovations including the development of a health transition plan and transition open days.

The health transition plan (popularly known as the passport document) helps identify the young person's transitional needs. The document assesses not just the patient's health needs, but the wider social, educational and developmental needs and gives the assessor (usually the transition nurse) the opportunity to discuss with the young person his/her hopes and aspirations. The Department of Health (2003, 2006) recommends that transition is a flexible process, allowing young people to explore their opportunities for independent living and develop skills in managing their condition, working towards readiness for the ultimate transfer to the adult facility. The transition passport, which allows needs to be re-assessed annually, progress to be noted, and areas of concern to be highlighted, provides this flexibility. The target age for transfer to the adult service is between 16 and 18 years but as the disease severity and cognitive impact of sickle cell disease can vary tremendously between individuals, planning transition must be a highly individualized, patient-centred and needs-focused process with age not necessarily an automatic trigger for transfer (Department of Health, 2006; Kennedy et al, 2007).

The transition clinic is held in the paediatric department, which is a familiar and non-threatening environment and this aims to reassure both the young person and his/her parents and to alleviate the fear that the paediatricians are abandoning them. The patients, parents, and paediatric and adult providers meet at this clinic to discuss a transition plan, and in this way the adult providers are perceived as positive additions to an already successful team (National Institutes of Health, 2002). This is a multidisciplinary clinic, and is attended by adult and paediatric members of the medical, nursing and psychology teams. Although the transition nurse and psychologist continue to support the young person in his/her first few years in the adult service, he/she is introduced to the adult nursing and psychology staff at this point. Sickle cell disease, like many chronic illnesses, can cause huge psychosocial problems and the ongoing support from the psychology team is invaluable in developing coping strategies. There is a well-established psychology team which is integrated into the adult sickle team and one of the psychology team facilitates a patient support group. This provides a forum for peer support and interaction for families. This includes input from the primary care team, community sickle nurses and counsellors who work closely with families in their homes.

During transition, parents often need just as much support as the young people. For most parents, the transition period signifies a loss of responsibility for, and control over, their child's medical care. They have invested so much into their children's care over the years that they may have difficulty relinquishing the central role, and this can produce resistance to the transition effort (National Institutes of Health, 2002). To address this, transition open days are held twice a year for young people and their families. The objectives for these events are to identify and understand the concerns, expectations, and preparation needs for the young people and their parents as well as to describe the perceived differences between the paediatric and adult care. The young people and their families also have the opportunity to visit the adult facility and meet their prospective adult carers and the day ends with an evaluation. The information gathered from these events has been useful in developing age-appropriate services in the adult facility, ensuring a seamless transition.

Role of the transition nurse and the psychologist

The transition nurse and psychologist are introduced to the young person between the ages of 12 and 14 years (depending on his/her cognitive and emotional maturity) and work with him/her through transition into the adult service until the age of 24 years, acknowledging that transition is a process that continues into adult services. The first meeting takes place predominantly in the paediatric consultant-led outpatient clinic but other opportunities include during acute episodes of hospitalization, in the day unit or in the adult outpatient clinic. By virtue of the cross-boundary working model, the clinical nurse specialist and psychologist provide the adolescents with a known and trusted professional to whom they can express their anxieties and provide a vital link between adult and paediatric services.

The role of the clinical nurse specialist is in part is to provide ongoing education to the adolescents to enable them to develop an increasing understanding of their condition and its implications, as well as to give more time to reinforce treatment plans which is not usually practical within a time-constrained clinic. Patients in transition may disengage from the service and a key role of the clinical nurse specialist involves helping them to coordinate clinic visits with work or college commitments and reminding them to attend clinic and investigations. Young people respond well to communication via mobile phone or email, therefore the nurse has a mobile phone which is used as a first port of call for patient queries. This has resulted in significant improvements in compliance.

The first admission to the adult hospital is a difficult and traumatic time for the young person with sickle cell disease. He/she moves from the secure, familiar environment of the paediatric hospital to the unfamiliar, busy adult environment, where he/she may be several decades younger than surrounding patients. The patient has to play a far more active role in his/her care and must be able to request analgesia as required. Support from the clinical nurse specialist is crucial during this time and includes spending time helping the patient adjust to a more proactive management style and in explaining differences in the two systems.

Having a dedicated psychologist integrated into the service and accessible to patients in hospital and in the community is essential in meeting a variety of clients' needs, as shown in *Table 2*. Follow-up psychological support and liaison with education, work or social services is offered by the transition team where necessary.

The frontal lobes are particularly sensitive to cerebrovascular disease in patients with sickle cell disease (Kral et al, 2001) and difficulties in executive functioning (e.g. planning, problem solving, decision making) often become increasingly apparent during adolescence, which has implications for the development of skills necessary for successful transition. The psychologist offers neuropsychological assessment to the adolescent and this is followed by written and verbal feedback and recommendations around appropriate support in education and implications for transition are shared with the team. Follow-up support is offered to strengthen planning, organizational and problem-solving skills.

The Department of Health (2006) suggests it is important to reinforce positive attitudes towards education and work. Accordingly, an information booklet has been developed about coping with sickle cell disease in higher education. This provides information about support networks, barriers and solutions for keeping well, advantages of open communication with friends and tutors and tips for positive communication. A coping plan for managing a crisis and catching up with work is also suggested. Additionally, a group to support young people with the transition to university has been developed. The group brainstormed strategies for coping on their own, keeping up with their studies and communicating about their condition, following modelling of problem solving, communication and assertiveness skills by the facilitators. Feedback suggests that these interventions are valued. Follow-up telephone counselling was requested by several young people which has enabled the psychologist to offer ongoing support, regardless of where young people are attending university. The young people were all happy to share numbers and email addresses and it is hoped that they will continue to use each other as a source of support.

Despite recent changes, the search for improvement continues by ongoing service evaluation and considering of further developments including the provision of separate adolescent in patient facilities.

Conclusions

The impact of a transition programme for chronic illness is deemed successful when the care of the chronically ill patient is transferred from paediatric to adult services in a seamless manner and when the adolescents are able to engage with professionals in the process. At an age where adolescents are taking more responsibility and planning their future, it is important to help them understand the effect of the illness on their life. The need to maintain good liaison between the adult and paediatric services can only be met by effective communication through the multidisciplinary team. Anxieties about the transition process can be mitigated in part by providing a comprehensive psychology service.

The role of the expert patient is enhanced by facilitating interaction between those about to move to adult-oriented care and those who have successfully achieved their full independence. This is invaluable, and if successfully done will equip the young people for adult life and ensure long-term engagement with adult health services.

Table 2. Ways in which the transition psychologist can meet the needs of the adolescent

Identification of barriers and solutions to keeping well
Support with transition to college or university
Stress management
Pain management
Sleep difficulties
Development of a positive identity
Confidence in communication about condition with peers or teachers
Communication within the family about management of condition
Neuropsychological assessment

KEY POINTS

- Sickle cell disease is the most common inherited disease in the UK, and with increased life expectancy the majority of patients survive to adulthood, putting greater burden on transition services.
- Transition is a challenging time for patients with sickle cell disease and can be associated with increased depression and anxiety, social isolation and lack of engagement with medical services.
- Improved preparation for and support during transition leads to improved self efficacy and fewer physical and psychological symptoms.
- A multidisciplinary team including medical staff, clinical nurse specialists and psychologists can be used to provide transitional care.
- A comprehensive transition programme could include a transition clinic, a health transition plan and transition open days.
- The role of a clinical nurse specialist includes education, coordination of care and support in the outpatient and inpatient environments.
- The role of a psychologist includes support with stress and pain management, support with transition to college or university and help with communication with family, peers and teachers.

There are few examples of transition programmes for young people with sickle cell disease in the literature, and this article describes the authors' local programme which has built on experience gained through evaluation of evidence-based programmes, implementing changes and reviewing the outcomes The lessons learnt from this model highlight the need for further research and formal evaluation of transition programmes. **BJHM**

Conflict of interest: none.

Anie K, Telfair J, Sickle Cell Disease Transition Study Working Group (2005) Multi-site study of transition in adolescents with sickle cell disease in the United Kingdom and the United States. *Int J Adolesc Med Health* **17**(2): 169–78

Blum RW, Garell D, Hodgman CH, Jorissen TW, Okinow NA, Orr DP, Slap GB (1993) Transition from child-centered to adult health-care systems for adolescents with chronic conditions. A position paper of the Society for Adolescent Medicine. *J Adolesc Health* **14:** 570–6

Bryant R, Walsh T (2009) Transition of the chronically ill youth with hemoglobinopathy to adult health care: an integrative review of the literature. *J Pediatr Health Care* **23**(1): 37–48

Burlew K, Telfair J, Colangelo L, Wright EC (2000) Becoming an adolescent; psychosocial factors. *J Pediatr Psychol* **25**(5): 287–99

Clay OJ, Telfair J (2007) Evaluation of a disease-specific self efficacy instrument in adolescents with sickle cell disease and its relationship to adjustment. *Child Neuropsychol* **13:** 188–203

Department of Health (2003) *Getting the right start: The National Service Framework for children, young people and maternity services. Part 1: Standard for hospital services.* Department of Health, London (www.dh.gov.uk/en/Publicationsandstatistics/Publications/PublicationsPolicyAndGuidance/DH_4006182 accessed 6 May 2010)

Department of Health (2006) *Transition: Getting it right for young people. Improving the transition of young people with long term conditions from children's to adult health services.* Department of Health, London

Doulton DM (2009) From cradle to commencement: transitioning pediatric sickle cell disease patients to adult providers. *J Pediatr Nurs* **25**(5): 287–99

Hauser ES, Dorn L (1999) Transitioning adolescents with sickle cell disease to adult centred care. *Pediatr Nurs* **25:** 479–89

Jones H, Turner C, Appleby H, Inusa B (2007) Transition services in sickle cell disease: Patients fears and expectations. Poster presentation. British Society of Haematology 47th annual scientific meeting: 30 April–2 May

Kennedy A, Sloman F, Douglass JA, Sawyer SM (2007) Young people with chronic illness: the approach to transition. *International Medicine Journal* **37**(8): 555–60

Kral MC, Brown RN, Hynd GW (2001) Neuropsychological aspects of pediatric sickle cell diesase. *Neuropsychol Rev* **11**(4): 179–96

Lavigne JV, Faire-Routman J (1992) Psychological adjustment to pediatric physical disorders: a meta-analysis review. *J Paediatr Psychol* **26:** 252–75

McPherson M, Thaniel L, Minniti C (2009) Transition of patients with sickle cell disease from pediatric to adult care. Assessing patient readiness. *Pediatr Blood Cancer* **52:** 838–41

National Institutes of Health (2002) *The Management of Sickle Cell Disease.* 4th edn. NIH Publication No 02-2117. www.nhlbi.nih.gov/health/prof/blood/sickle/sc_mngt.pdf (accessed 6 May 2010)

NHS Sickle Cell and Thalassaemia Screening Programme (2006) *Sickle cell disease in childhood. Standards and guidelines for clinical care.* NHS, London (sct.screening.nhs.uk/cms.php?folder=2493 accessed 6 May 2010)

Okpala I, Thomas V, Westerdale N et al (2002) The comprehensive care of sickle cell disease. *Eur J Haematol* **68**(3): 157–62

Platt OS, Brambilla DJ, Rosse WF, Milner PF, Castro O, Steinberg MH, Klug PP (1994) Mortality in sickle cell disease. Life expectancy and risk factors for early death. *N Engl J Med* **330:** 1639–44

Ricker JH, Delamater AM, Hsu J (1998) Correlates of regimen adherence in cystic fibrosis. *J Clin Psychol Med Settings* **5:** 159–72

Sickle Cell Society (2008) *Standards for the Clinical Care of Adults with Sickle Cell Disease in the UK.* Sickle Cell Society, London www.sicklecellsociety.org/pdf/CareBook.pdf (accessed 6 May 2010)

Streetly A, Latinovic R, Hall K, Henthorn J (2009) Implementation of universal newborn bloodspot screening for sickle cell disease and other clinically significant haemoglobinopathies in England: screening results for 2005-7. *J Clin Pathol* **62**(1): 26–30

Telfair J, Myers J, Drezner S (1994) Transfer as a component of the transition of adolescent with sickle cell disease to adult care: Adolescent, adult and parent perspectives. *J Adolesc Health* **15:** 558–65

Telfer P, Coen P, Chakravorty S et al (2007) Clinical outcomes in children with sickle cell disease living in England: a neonatal cohort in East London. *Haematologica* **92**(7): 905–12

Tuchman LK, Slap GB, Britto MT (2008) Transition to adult care: experiences and expectations of adolescents with a chronic illness. *Child Care Health Dev* **34**(5): 557–63

Watson AR (2000) Non-compliance and transfer from Paediatrics to adult transplant unit. *Paediatric Nephrology Journal* **14:** 469–72

Wierenga KJJ, Hambleton IR, Lewis NA (2001) Survival estimates from patients with homozygous sickle cell disease in Jamaica: a clinic-based population study. *Lancet* **357:** 680–3

Williams PG, Holmbeck GN, Greenley R (2002) Adolescent health psychology. *J Consult Clin Psychol* **70**(3): 828–42

Wojciechowski EA, Hurtig A, Dorn L (2002) A natural history study of adolescents and young adults with sickle cell disease as they transfer to adult care: A need for case management services. *J Pediatr Nurs* **17:** 18–27