

Transition to Adulthood for Young Men with Duchenne Muscular Dystrophy

Duchenne Muscular Dystrophy (DMD) is an inherited neuromuscular disease which affects boys. During the last few years there have been significant improvements in the ways DMD is managed. As a result, young men with DMD are now living longer than ever before and face a particular set of challenges as they reach adulthood. Those who are the focus of this study, aged over 15 years old, have reached, or are reaching stages of adulthood that were, at the time of their childhood, largely unexpected and unplanned. Given increases in life expectancy, young people with DMD should be looking forward to living independently, with appropriate support, as adults.

The key objectives of this study were:

1. To investigate, from their own perspectives, how the health and well-being of young men living with DMD, and that of their parents, can be maximised, particularly at the transition to adulthood.
2. To consider the potential contribution of the National Service Framework for Long Term Neurological Conditions for this group of people.

The research was carried out by David Abbott and John Carpenter at the School for Policy Studies (University of Bristol) and in partnership with the International Centre for Life (University of Newcastle), the Duchenne Family Support Group, the Muscular Dystrophy Campaign, and with support from Action Duchenne. The data were collected between October 2007 and November 2008.

The research comprised:

1. A postal survey of parents with a son with DMD aged 15+ living in the South West, the West Midlands and the North East of England.
2. Face to face interviews with 40 young men, their parents and siblings about growing older with DMD and the issues they faced at transition.

Findings

1. Postal Survey

A third of the parents living in the three regions, 38 out of 121, mainly mothers, completed an anonymous postal questionnaire.

Family circumstances

Eight in ten of the young men with DMD were living in a family with married or cohabiting parents. Most mothers reported a high level of support from their partners however over 80% reported clinical levels of anxiety and depression. Most had been woken by their son every night of the previous week (average 4.6 nights a week). Half were looking after the family full time, but over a third were in full or part time paid employment.

Disposable family income was comparable to the general population, but this does not take account of the three times greater additional expenditure incurred in looking after a disabled young person. Take up of social security benefits was high but only a quarter were using a direct payment. Two thirds owned their own homes and most considered their accommodation suitable.

All families had their own car or van, in most cases adapted. Travel by public transport was difficult; in two thirds of families, the young man's pain and discomfort was a significant obstacle to going out.

Over half of parents were able to have a break from caring from their son as often as needed; in most cases when their son stayed in a hospice. Some did not get sufficient breaks and others had chosen not to use them. Around a quarter were using, or had used a support group; half did not use a support group by choice.

Services

Most families had been in contact with a large number of professionals in the previous six months. Up to half had been to a specialist medical clinic including respiratory and cardiac clinics and muscle services. However, only a quarter had a key worker or care coordinator, usually a social worker.

Overall, half the parents recalled that some form of transition planning had taken place at school, but over a third did not think this had happened. In most cases, parents considered that their

sons had played an active part in planning his future. Two thirds of parents had insufficient information about options for what their son did next in his life, the services that are available to support him, the roles and responsibilities of professionals and support available for them as parents.

2. Interviews with young men and their families

Education, Work and Beyond

Half the young men interviewed were, or had been, in a mainstream school, although their families sometimes had difficulty in persuading a school that they could meet their son's needs. Positive experiences of school included particularly helpful teachers or other staff; involvement in out of and after school activities; and schools with accessible buildings and on-site services such as hydrotherapy. Negative accounts featured bullying, difficulties with transport; some "difficult" Special Education Needs Coordinators; problems with accessing buildings and thus the full school curriculum; experiences of being excluded from activities outside and after school.

Most families were unclear about the process of formal transition planning (required by government guidelines); key professionals were often absent and there were few examples of young people being actively involved. Parents felt the onus was on them to, 'sort things out.'

Almost all saw college as the next step, although a few were not sure they really wanted to go. There was often not very much choice of college because of access problems; some were directed towards residential college.

Most of the young men were positive about college, saying it gave them the opportunity to inhabit more of an adult environment and make friends. However, some found making friends harder at college; some felt that their academic capabilities were not taken seriously and that staff did not have enough understanding and expertise about disability in general and DMD in particular. Problems of access to college buildings and out of college activities effectively barred them from some subjects, social events or fieldtrips.

Most young people remembered getting advice about what they should do next, but this was often insufficiently specific about local options. Some gave up on college when they got tired of having to deal with access problems.

Five were at, or had been at university. This had involved much planning. Experiences were positive, but there were difficulties in organising reliable and sustainable care and support

Only one person had had a paid job. Those who had tried to find jobs faced insurmountable difficulties related to employers' attitudes, access problems, and a lack of specialist and useful advice. The older young men in the study thought they were unlikely to get jobs and that they were probably not well enough to work.

At the time of the interviews, about a third of the young men had been at home during the day and not doing any kind of education, training, or work for between few months and seven years. Their social lives were very restricted (see below).

Families' experiences of services

All but three of the families characterised their overall experience of services as problematic. Getting their son's needs met involved repeated 'battles.' Young men with DMD were often aware of these disputes and found them upsetting and stressful. There were very few examples of professionals acting as effective care coordinators for families, who felt that most things were down to them to sort out. The most common problems concerned: the provision of care/support packages; decisions about residential education; obtaining equipment including wheelchairs; and housing and adaptations.

Those families which were positive about services in general stressed factors such as good coordination (through a muscle clinic), planning ahead, and long standing relationships between the family, the young man, and helpful professionals.

Almost all families in the North East attended a Muscle Centre where staff's specialist knowledge and expertise were greatly appreciated. The Centre was available for young men of any age, with no transfer to adult services; this was greatly valued. In the West Midlands, most families saw a consultant paediatrician in

childhood, about whom they were very positive. However, some families were unsure about transfer to adult services and about whom they would be seeing and why. Others said that they were given a lot of notice and that there was some flexibility. In the South West, there were no adult neuromuscular consultants. Families were confused and anxious about transition pathways; a few relied very much on their GPS.

Many parents and young men expressed disquiet with the way that appointments in later childhood and early adulthood focused on charting their deteriorating health; this was demotivating even if clinicians thought it was important to monitor health status.

Physiotherapy was as one of the first and most significant losses following their transition to adult services. There was wide variation in the support obtained from occupational in relation to equipment and adaptations.

Just under half the families had had contact with children's social care and most of these were critical of the nature and level of support they received. However others said their social worker was very supportive. Social workers' involvement at transition tended to focus on paperwork and meetings rather than any outcomes or support. Following transition to adult social care, there was no on-going relationship with a named social worker; instead, they were dealt with by a duty social worker.

Most families used their local hospice for short breaks and were overwhelmingly positive about the nature and quality of support they experienced. As they grew older, some young people were no longer able to attend their local hospice and in some cases there was no suitable alternative.

In two of the three regions family care officers/neuromuscular care advisors provided information and support; this was highly regarded.

Young people's social lives, independence and support

Young men at school or college generally had adequate social lives and enough friends. However, most who had finished education were leading restricted lives. TV, computer games and

going out with close family members took occupied their time but most said that they would like to do more. Parents had mixed views: some were worried that their son's potential was being wasted but were anxious not to pressurise them. The main obstacles to doing more during the day were a lack of information about local options which took account of the young person's health and impairments, their transport and support needs, and the availability of other people to do things with. It was hard to gain independence from their parents because they relied on them for transport and care. Few families were using direct payments to pay personal assistants to support their son's leisure activities.

Four young men at college had girlfriends; this was easier for those living away from home. Others, and their parents, wanted a relationship but worried that it might be difficult to find a partner.

Support and care

External support for the young men varied from a couple of hours a day to a 24 hour package of care, even for those with similar levels of need. Most parents were still very actively providing care. It could be difficult for parents and their sons to have enough physical and psychological space from each other. Some families, concerned for privacy, declined external carers. The quality of external care varied enormously and its inflexibility could impede the young person's independence.

About one third of parents were using a direct payment. Recruiting reliable carers was difficult. Some were confused about their entitlement and how they could be used. Others felt 'pushed into' direct payments and found the organisational responsibilities onerous.

Living with DMD and becoming an adult

Families dealt with DMD in a whole range of ways which changed in response to events such as a decline in health, a major medical intervention, or the death of friends with DMD. Young men tended *not* to think about living with DMD, although it was always at the back of their minds. It was hard to stay positive if they thought about it too much. However some valued the chance to talk to people they trusted. Parents tended to resist outsiders suggesting that they should, or *how* they should, talk about DMD and its consequences.

Some parents suggested that their sons tried to protect them by not talking or showing their emotions. Nonetheless, young men identified their parents as their stop for questions or concerns. Parents were not always sure what their son knew about DMD; they were unsure what to say given changing medical technology and increases in life expectancy. A small number of families discussed their emotional needs in relation to living with DMD.

Families tended to say that they 'lived for the day' and found it difficult to look very far ahead because they felt uncertain and apprehensive. But they *did* plan ahead – for holidays, what to do once school or college was finished, and how to organise care in the future. Parents struggled with deciding how much pressure to put on their sons to get qualifications, think about work and living away from home, and with expectations for their futures. Some felt their son's life expectancy was such that there was no point in having expectations; some parents thought that professionals used this to ration services. There were very strong emotional bonds between parents and their sons, mutually supportive and often characterised by warmth and humour. However, this closeness meant that some young men did not feel able to develop independence – a concern shared equally by some parents.

DMD, however significant, was just one aspect of family lives. Households had other siblings, busy work lives, domestic work, socialising, and all the ordinary business of family life. However, some said that DMD dominated family life – appointments with professionals, fighting for services, caring responsibilities and so on. In addition, parents disliked professionals treating their son as if he had no identity other than DMD. Some parents got involved in support groups and with other families affected by DMD; some did the opposite, choosing not to get involved because they did not want the frequent reminders of living with DMD.

Young men with DMD were adversely affected by society's disabling barriers such as poor physical access and discriminatory attitudes, and services which did not meet their needs. In addition, the physiological affects of DMD restricted their independence. Pain was frequently mentioned as a source of distress. Medical interventions such as spinal surgery could dramatically affect their lives.

Many of the findings of this study are directly applicable to a much group of young disabled people, with long term conditions, with complex health care needs, and with life limiting illnesses. It confirms previous research about transition to adulthood for young people and the very many obstacles they face. It shows that while advances in health care have increased life expectancy, this has not been matched by an increase in support at home and in the community. The challenge posed by young men with DMD and their families is to ensure that their shorter lives are no less valuable and just as fulfilling as any others in our society.

The report concludes by considering the findings in relation to the Quality Requirements of the National Service Framework (NSF) for Long Term Neurological Conditions (2005) and its potential to improve the lives of people with DMD and their families.