The adolescent with disability

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Everything is there when they're children - it's handed to you on a plate, but suddenly they get to nineteen and everything disappears - they get discharged from hospitals and then it's only if you have a problem that you can get a check up ...I mean, suddenly, they get better?

Mother of profoundly disabled young man

The term 'disability' refers to:

any restriction or lack of ability to perform activities in the range considered normal for a human being.

The impact of disability (the resulting handicap or disadvantage) varies with changing life situations, and depends on the attitudes of society and the extent of environmental adaptations (eg access to buildings for wheelchair users). Any chronic illness can be disabling, but the term 'disability' is most commonly applied to neurological and orthopaedic conditions, especially those shown in Table 1.

The principles of care for disabled adolescents are the same as those for other long-term medical problems in this age group, but some additional issues require special consideration. These centre around the decreased ability of disabled young people to achieve the main task of adolescence, the attainment of physical and emotional independence.

Variable severity of disability

The conditions listed in Table 1 vary widely in severity. For example, mild hemiplegic cerebral palsy or mild to moderate spastic diplegia may allow a young person to achieve an independent adult life, perhaps restricted only by an inability to participate in sports requiring speed and coordination. At the other extreme, a young person with severe four-limb cerebral palsy associated with learning difficulties, cortical visual impairment and epilepsy will require lifetime total care.

The need for autonomy

As children enter their teens, they seek greater autonomy in peer group interactions and sexuality. Their need and right to take increasing responsibility for

Key Points

- Disabled adolescents have difficulties in developing autonomy and independence
- Severely disabled children surviving to adult life have complex medical care needs
- Parents are the main carers for disabled adolescents
- There is no coherent service provision for this forgotten group
their own health are recognised in the concept of Gillick competence. This originated in a legal challenge by the mother of a teenage girl to medical professionals as to their right to provide contraception to a girl below the age of 16. The case went to the House of Lords. The wise judgement of their Lordships was that a person acquires increasing responsibility for his or her own health by an evolving process during adolescence, not by an overnight transformation at the age of 16. This places the onus on the professionals to consider the extent to which young people are competent to understand issues to do with their own health and health care. The acquisition of this autonomy is often delayed in disabled young people, partly because of their impaired ability to reason and to communicate their views, but also because their carers fail to perceive their need to develop independence. Carers unwittingly reward passivity, so that disabled young people continue allowing others to make decisions on their behalf. Thus, the protective instincts of parents and professionals, so necessary for the survival of severely disabled young children, can be a negative influence for the disabled teenager. The issues which interest healthy youngsters - teen culture, sexuality, relationships, sport, career ambition - are often a closed book to disabled teenagers. They cannot go out alone, and they depend on parents for their social life. Loneliness is a common complaint for those able to articulate their emotions.

The care of disabled young people

For young people with disabilities that preclude an independent lifestyle, there are few options. The optimum level of independence can often be achieved in a residential school placement, followed by accommodation in, for example, a community or sheltered housing development. Places in these schemes are seldom sufficient for the demand, and their high cost is a disincentive to social services and beyond the reach of most families. The usual result is that disabled young people stay at home with their parents or, more often in recent times, with a single or unsupported parent. Three demographic trends are important in this context:

- mothers are having their children at an older age
- severely and profoundly disabled children are living into their 30s and beyond
- the lifespan of the parents is lengthening.

Together, these generate the prediction of elderly lone parents, perhaps 70 or 80 years old, still caring for profoundly disabled offspring. The needs of severely disabled people vary widely. Paradoxically, parents may find it relatively easy to care for a person with very severe four-limb cerebral palsy, severe learning problems and very little insight. Such individuals may be easy to lift and carry because they are underweight and undernourished due to swallowing difficulties associated with pseudobulbar palsy and to the effect of brain damage on growth. Of course, feeding and bathing them, and dealing with their incontinence are tiring, repetitive tasks, but carers can manage their daily routines around the disabled person. Social service care teams and home adaptations may lighten the burden.

In contrast, the young person with severe learning problems or autism may be fully mobile and physically capable, but exhibit a range of unpredictable and socially undesirable behaviours which demand constant supervision by day and night, sometimes even physical restraint, leading to considerable stress and a complete loss of any other social contact for the carer.

Evolving disorders

Much disability is due to a condition in which the basic pathology is static, but there are nevertheless many new medical problems which may need attention. Epilepsy may change in pattern or severity (see the accompanying article by Cross). Aethaloid cerebral palsy may cause a range of orthopaedic and neurological degenerative signs and symptoms, associated in particular with the incessant neck movements. Reflux oesophagitis is common in cerebral palsy. It may get worse when the disabled individual is constantly at home and no longer benefits from the frequent position changes and stretching supervised by physiotherapists at school. Stress, frustration and anxiety probably make matters worse. Other examples of medical complications occurring in disabled people include:

- deterioration in unstable hip joints and scoliosis
- the appearance of new neurological signs associated with neural tube defects due to cord tethering
- sudden life-threatening malfunction of a shunt valve in hydrocephalus
- urinary tract infections.

Those with a chronic disabling condition are of course not immune from the full range of other medical and surgical

**Table 1. Neurological and orthopaedic conditions to which the term 'disability' is most commonly applied.**

| Condition | Description |
|-----------|-------------|
| Cerebral palsy | Often associated with hydrocephalus |
| Muscle disorders | (e.g., Duchenne muscular dystrophy) |
| Acquired brain injury | (trauma, CNS infection) |
| Severe learning difficulties | (previously called severe mental handicap) |
| Moderate learning difficulties | (previously called mild/moderate mental handicap) |
| Specific language impairment | (childhood dysphasias) |
| Autism and autistic spectrum disorders | |
| Permanent congenital hearing impairment | |
| Visual impairment | |
| Epilepsies | |
| Osteogenesis imperfecta | |
| Bone dysplasias | |
| Multisystem syndromes | (e.g., neurofibromatosis type 1) |

CNS = central nervous system
disorders afflicting the population at large, but diagnosis may be difficult in people who cannot easily communicate. Psychological and psychiatric disorders are common, particularly in those with severe communication disorders such as deafness or dysphasia, but for obvious reasons present a particular challenge in diagnosis. Strange behaviours in people with learning difficulties may too readily be attributed to epilepsy, but often an underlying psychological explanation can be found and is sometimes rewardingly simple to address.

Some disabling childhood conditions are progressive and deteriorate over time. For example, most boys with Duchenne muscular dystrophy die in their late teens or early 20s. They are often cared for throughout their lives by paediatricians, as handover to adult services just a few years before their death does not seem sensible. There are other less common disorders which deteriorate: for example, Rett's syndrome and a wide range of metabolic and storage disorders. Sometimes mobility is lost during the teens due, for example, to growth and weight gain, or to a progressive neurological disorder that had been mistaken for cerebral palsy (which by definition is due to a static lesion), but in many cases the reasons are not clear. Some multisystem syndromes are associated with well recognised risks of new problems emerging, and careful follow-up is vitally important. The best known examples are neurofibromatosis and tuberous sclerosis.

Handover – benefits of the team approach

Most disabled children and younger adolescents are managed with a multidisciplinary team approach, based in a child development centre. Paediatricians differ in their approach to handover to adult services. Some continue to offer care 'unofficially' until the young persons are in their late teens or even their 20s, especially when there is a deteriorating or complex disorder. Others strictly hand over or discharge at a fixed age. Much depends on whether or not there is an adult service to receive them.

The transition plan

Proper planning of the transition process is vital. The 1994 Code of Practice associated with the Education Act 1981 and subsequent legislation defines the duties of the education authority for children who have a Statement of Special Educational Need, and specifies that at the age of 14 they should have a detailed review. This provides an opportunity for professionals in health, education and social services to plan with the young person and the family what will be needed when he or she leaves school. This opportunity is not always well used; furthermore, it applies only to young people who have a Statement. It is possible for an individual with normal intellect but severe physical and mobility problems to cope adequately in school without having a Statement. There is no statutory duty within the Code to these young people, but they would also benefit from a review.

The importance of primary care

It is particularly important that the primary health care team, and the general practitioner (GP) in particular, are involved in this process. There is a tendency for paediatricians to provide not only specialist care for their disabled child patients but also to respond to problems that would normally be the province of primary care. This can result in the GP hardly knowing the child, and it is unreasonable to expect that he or she should then suddenly assume full responsibility for that individual's care without adequate briefing. One simple way of addressing this problem is to ensure that both parents and the primary care team receive complete copies of all assessments and reports, including the one produced at the 14-year old assessment. The report should not only describe the current problems, but also list possible problems that might arise and who might best deal with them.

Which specialty caters for disabled young people?

Ideally, the transition process should involve handover to a named consultant team which can provide the same multidisciplinary cover as that offered in paediatric practice. This is particularly important for neural tube defects and other complex disorders, but sadly still rare. Several models of care exist, but none is completely satisfactory or widespread. A 'nurse for transition' can help the family work out what arrangements would be most suitable for the teenager, and undertake the necessary liaison. A few paediatricians offer a clinic service for the young people who have been their patients, but in a different setting to emphasise their increasing autonomy and need for an adult- rather than a child-centred clinic environment. Others have set up a joint clinic with a consultant in neurology or rehabilitation medicine where young people can be seen and problems jointly resolved. Subsequently, care is handed over to a single consultant who can act as the focal point, most usually a consultant in rehabilitation medicine, a neurologist, psychiatrist or consultant in learning difficulties.

One alternative is for the GP to supervise the health care of disabled individuals. This could be a logical approach, but few GPs currently feel they have had adequate training for this task, and have difficulty in deciding which specialist to refer to, given the often undifferentiated nature of the complaints presented by severely disabled people. Preventive health care for severely disabled people (eg dental care, breast and cytology screening) is unsatisfactory. Even more difficult is the prevention of the social ills to which these individuals are prone: for instance, they are at considerably increased risk of physical and sexual abuse.

Conclusion

There is as yet no satisfactory model for the handover of the medical care of disabled adolescents. Several alternatives
exist, but none has been adopted with any enthusiasm except in a few districts where excellent services have been created. The issue should be jointly addressed between the medical and nursing colleges and the social care agencies.

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Epilepsy and the adolescent

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Life poses many difficulties for the individual approaching adolescence, even without the additional burden of a diagnosis of epilepsy. For many, epilepsy will have been diagnosed earlier in childhood, and the taking of medication already be a daily ritual. For others, the diagnosis may be made at this time, and a challenge presented as to how this may now affect their lifestyle. Whatever the situation, it has to be remembered that this time brings much in the way of emotional and physical changes which may not only affect the seizure pattern but also raise questions that need to be addressed as the individual approaches adulthood. It is a time when individuals need to feel in control, and all discussions have to be undertaken with this in mind.

Diagnosis and prognosis

The importance of ensuring that a correct diagnosis is made is paramount, whether an individual presents with ongoing seizures that appear resistant to medication, or as a new presentation with apparent epileptic seizures. Continued reassessment and consideration of alternative diagnoses are required, remembering that the rate of false diagnosis of epilepsy could be at least 10% (Table 1). The diagnosis of the epilepsy syndrome is also important, to maximise prognostic information and outline the likelihood of long term medication to both parents and the teenager. Diagnosis of the syndrome identifies those in whom epilepsy is likely to persist into adulthood, and presents the possibility of alternative management such as surgery (Table 2). In addition, any implications of the underlying aetiology (eg genetic counselling and regular neuro/renal imaging in tuberous sclerosis) need to be fully understood.

Medication

Depending on the age when their diagnosis was made, teenagers are likely to have grown up with decisions usually made by parents. Even if they are seizure-free, the issue of ongoing medication provides them with a different identity to their peers. It is necessary for a teenager to take a role in this as early as possible, with self-documentation of seizure frequency (by the use of

Table 1. Differential diagnosis of seizures in adolescence.

| Symptomatic and related disorders | Vasovagal |
|----------------------------------|-----------|
| Hyperventilation                 | Cardiac   |
| Behavioural/psychiatric          | Anxiety related |
| Psychotic episodes               | Pseudoseizures |
| Pseudoseizures                   | Ritualistic movements |
| Neurological                     | Migraine |
| Sleep phenomena                  | Tics      |
| Intermittent and recurrent ataxia| Paroxysmal dystonias |
| Paroxysmal dystonias             | Transient ischaemic attacks |
| Sleep jerks                      | Sleep walking |
| Sleep walking                    | Narcolepsy |