EDITORIAL

Adolescent neurology

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Clinicians taking over teenagers' care must not only assess their neurological condition, but also appreciate the influence of evolving physical, emotional, educational and social needs on their predicament.

INTRODUCTION

Adolescence, Erikson's fifth stage of man (Erikson 1965), is the phase of identity and role confusion. Teenage angst was epitomised by Adrian Mole whose analysis of his physical changes, acne, relationships, self-identity and growing independence in society, repeatedly clashed with his legal position as a child (Townsend 1982). Normal adolescence itself presents major challenges, but teenagers' problems are greatly exacerbated by the additional burden of a neurological disorder. Their clinical care falls between paediatric and adult services, and their special problems can go unrecognized. Adult neurologists may see teenagers presenting either with new problems, or passed on from paediatric care with chronic conditions.

TRANSITION

The recent UK National Service Framework for Children, Young People and Maternity Services, identified arrangements for transition from paediatric to adult care as crucial to successful continuity and effectiveness of teenagers' clinical management (Craft 2003). Transition is not just an administrative event – it is a guided process of educational and therapeutic transfer to adult care, and should be planned, purposeful, co-ordinated, uninterrupted and developmentally appropriate (Tuffrey & Pearce 2003). Box 1 summarizes some of the problems.

Disabled children increasingly survive to adulthood and 90% now live beyond the age of 20 (Blum 1995). Epilepsy, cerebral palsy and conditions with learning disability are the most common reasons for follow-up in paediatric neurology clinics. Many have untreated health problems, compounded by reduced social and health care contact after leaving school. Transition must minimize adverse physical and psy-

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chological outcomes, and avoid dropout from healthcare services. The disabled teenagers most satisfied with their service provision have come through clearly defined handover processes (Fiorentino *et al*. 1998).

Focused transition clinics, specialty clinics, and primary care controlled services can assist the process (Callahan *et al*. 2001), but with little evidence to identify the best model. Adult and paediatric neurologists in joint transition clinics may be the gold standard, but are practical only for conditions with sufficient patient numbers, e.g. epilepsy.

Adult and paediatric specialties are culturally different. Paediatricians emphasize family centredness and working with education and community services. They may sometimes doubt the adult services' ability to provide for the teenager, and these feelings can be transmitted to the teenager, undermining confidence. Teenagers frequently have enjoyed long-standing relationships with paediatric teams, though often taking little active control of their own illness management. Both paediatric and adult teams must ease the teenager towards a more adult orientated style, and believe positively in the process.

Transition to adult services is sometimes developmentally inappropriate. Children with severe neurodegenerative or neuromuscular diseases surviving into adolescence may stay appropriately with the paediatric services. Inappropriate adherence to age-based paediatric admission policies can cause difficulties for 17–19 years olds; in an emergency, admission to an adult acute ward can be an abrupt disaster.

THE FIRST ADULT CONSULTATION

The initial consultation should focus on diagnosis, investigations and treatment, checking understanding, providing information, and fostering the new relationship. Young adults want clear explanations; treatment as an equal; a friendly, reassuring, confidential approach; and time taken to listen (Lansdown 2000).

Diagnosis

Appropriate clinical care invariably requires the correct diagnosis. Even static conditions, such as cerebral palsy, justify re-evaluation and consideration of re-investigation. However, over-enthusiastic questioning of the patient's history and diagnosis (as the initial approach) risks unsettling patients and parents after years with a trusted paediatrician.

Investigations

Recent imaging advances justify review of the adequacy of previous radiology. Genetic advances also merit consideration of further testing. Chromosomal abnormalities are increasingly implicated in the epilepsies, and subtelomeric deletions in the learning disabilities (De Vries *et al*. 2001). Metabolic screening is indicated for neurological disorders showing fluctuation, deterioration, or triggering by feeding or fasting. The review of Gray *et al*. (2000) and Phillipa Lamont's article in the next issue of *Practical Neurology* should help neurologists to 'think metabolic' (Lamont 2004).

BOX 1 PROBLEMS WITH TRANSITION OF TEENAGERS TO ADULT SERVICES

- Loss of long-standing relationship with paediatric services.
- Different clinic dynamic: more adult approach, more often seen alone, less frequent follow-up.
- Lack of equivalent role of community paediatricians in adult services.
- Handover is condition driven: referral to different subspecialty adult neurologists makes joint clinics with paediatricians difficult.
- Children with multiple problems may be attending several paediatric specialists.
- Specialist paediatricians may have under-used primary care providers, undermining the primary care team's subsequent relationship with the teenager.

Treatment

Drug licensing creates prescribing difficulties with two-thirds of hospitalized children receiving unlicensed or off-label medicines (Choonara 2000). Pain management epitomises the problem, with little evidence to guide clinical practice in children and adolescents (Eccleston & Malleson 2003). For example, only one triptan preparation is licensed for 12-year-olds, yet there are six to choose from when aged 18 years. Adolescents therefore grow into an evidence base more familiar to adult clinicians than paediatricians, making therapy review useful.

Information and understanding

The adult setting provides an opportunity to explore topics such as alcohol, driving, sex, contraception, and pregnancy. The last may be particularly important in teenagers with genetic disorders. Clinical nurse specialists are an invaluable information source for teenagers. Questionnaires and motivational interviewing may help to check teenagers' understanding, improve compliance, and reduce anxieties. Internet browsing is a teenager-acceptable way to foster knowledge and enhance illness control. Box 2 gives useful websites for patients and professionals. It is sensible to check a site before recommending it to a teenager, especially as only 1% of information on patient orientated websites is directed at children.

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BOX 2 ADOLESCENT NEUROLOGY: USEFUL WEB SITES

1 British Paediatric Neurology Association
Contains links to 2 and 3.
http://www.bpna.org.uk
2 Child Neurology Home Page.
Provides 'Family Village' library with information on neurological diagnoses.
http://www.waisman.wisc.edu/child-neuro/index.html
3 Online Mendelian Inheritance in Man (OMIM).
Search clinical features for syndromes, e.g. macro-orchidism gives Fragile X.
http://www.ncbi.nlm.nih.gov (click OMIM)
4 Neuromuscular homepage
http://www.neuro.wustl.edu/neuromuscular/
5 Charity: Contact a Family.
Support, information, and group contacts for many syndromes and disorders. Manual available.
http://www.cafamily.org.uk
6 Bridging the gaps: Healthcare for adolescents.
An intercollegiate working party report on adolescent health
http://www.rcpch.ac.uk/publications/recent_publications.html
7 National Service Framework for Children and Young People.
UK standards of care for people under the age of 19 years.
http://www.doh.gov.uk/nsf/children.htm
(Sites accessed 27 August 2003)

PRACTICAL CONSIDERATIONS

- Teenagers commonly deny or minimize their symptoms, through bravado, embarrassment, and to avoid being different. Their attitude to illness may also be surprising. The clinician must balance respect for the resilience and bravado of youth with the need to tease out denied problems, which may require intervention and support.
- Judging the appropriate parental contribution to the consultation is difficult. Teenagers with chronic conditions naturally retain strong attachments to parents who deliver (and often must continue to deliver) significant levels of care and emotional support. Although conflicting with their need for independence – inevitably already delayed in those with chronic disabilities – parental contributions and input must be respected. The consultation focus remains firmly on the teenager, but with the parents tactfully included.
- Time alone with the teenager is essential during the consultation. Time alone with the parent may also be revealing. History taking with parents present sometimes provokes only horror, silence, or monosyllables. Most teenagers blame embarrassment for concealing information from their health professional. A useful tactic is to perform the physical examination in a separate room, using the opportunity to complete the history taking away from parents.
- Self-conscious teenagers may be reluctant to undress for examination. Encouraging them to wear football/sports shorts beneath their clothing when attending clinics can avoid embarrassment.

MAJOR NEUROLOGICAL DISORDERS

Whilst almost any neurological disorder can present in adolescence, some have features peculiar to this age group. For example, Huntington's disease presents differently in teenagers (hypokinetic/rigid) than in adults. Certain conditions typically start in adolescence (migraine, nonepileptic attack disorder, juvenile myoclonic epilepsy), while others remit (benign partial epilepsy of childhood with centro-temporal spikes). Some pre-existing conditions become more recognizable following puberty, e.g. Fragile X syndrome, or characteristically show their first symptoms in the second decade, e.g. syringomyelia (Mariani *et al.* 1991).

Epilepsy

Epilepsy is the commonest neurological problem in patients transferred from paediatric to adult care. Teenagers with epilepsy present particular challenges because not only is their independence threatened, but also their behavioural response to the condition may have an adverse social effect. Furthermore, a condition that is paroxysmal is easier to deny.

Emotional, behavioural, and relationship difficulties occur in 40% of children with epilepsy, compared to 10% with diabetes mellitus or controls (Davies *et al.* 2003). This suggests that social stigma and associated neurological problems exaggerate the burden of the chronic condition. The high prevalence of epilepsy provides an incentive to develop teenage epilepsy clinics (Smith *et al.* 2002).

Neuropsychiatric and neurodegenerative disorders

Depression is commoner in adolescence than at other childhood stages, especially with an underlying chronic physical disease. Apparent (non-organic) regression may result from educational demands and additional pressures upon a teenager who hitherto had coped. Conversely, intellectual decline and psychiatric

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disorders may easily be overlooked or misdiagnosed through being initially wrongly attributed to extremes of normal adolescence.

Adult neurologists may, less readily than paediatric colleagues, consider late or unusual manifestations of paediatric conditions, e.g. metabolic disorders, as the cause of neurological regression. The UK surveillance programme for progressive intellectual and neurological deterioration (PIND) in childhood illustrated the scope of the problem in identifying 360 children and teenagers with 88 separate neurodegenerative disorders (Verity *et al*. 2000). Forsyth's article is helpful in planning neurodegenerative investigations (Forsyth 2003).

Neuromuscular disorders

One in 3000 people has a serious neuromuscular disorder, often beginning in childhood. This group comprises about one-quarter of those requiring planned transition. For those with major physical disability such as Duchenne muscular dystrophy, anything approaching normal adolescence (socially, sexually, etc.) is impossible, with remorseless progression during the teenage years. In practice, their greater dependence on carers, and long relationships with multidisciplinary staff members, often keeps such patients within the paediatric service.

SERVICE AND TRAINING DEVELOPMENT

Adolescent medicine is becoming a separate discipline in Australia and the USA (Callahan *et al*. 2001), and similar developments seem inevitable in the UK. However, this will first require closer working between paediatric and adult services, and changes to training. In the UK, trainee paediatric neurologists have 6 months compulsory training in adult neurology, and 3 months in child and adolescent psychiatry. By contrast, adult neurology training requires only nominal exposure to paediatric neurology, learning disabilities and neuropsychiatry, and no predefined training period.

CONCLUSIONS

The diversity of neurological presentations and diagnoses in teenagers, their social and developmental concerns, and the lack of evidence to support and inform practice, each presents major challenges. The neurological disorders encountered in teenagers are similar to those in other age groups. However, their problems are different and more pressing, because of their ongoing physical and social development, heightened self-awareness and anxiety, and low self-esteem. The resulting social, educational, and developmental handicaps, once established, may persist even with improvement of the neurological disorder.

There is some comfort in the temporary nature of adolescence. Yet even as Adrian Mole leaves behind his cappuccino years (Townsend 1999), so the beleaguered Harry Potter faces the additional trials of adolescence (Rowling 2003).

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