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 of children with complex pathology, and increasing prevalence of lifestyle-related conditions such as obesity, hypertension and type 2 diabetes. Specific factors that make transition planning important in young people with LTCs are outlined in Figure 31.1. Planning the process of transition from paediatric to adult health services and improving the assessment of young people as they enter those adult services have been shown to impact positively on long-term health outcomes. There is a need for physicians to gain new skills in the care of young people and adults who have conditions that have arisen in childhood. This includes developing specific skills in the management of adolescents and young adults, managing the process of transition and developing knowledge of relevant medical conditions. The overall approach to transition medicine, as well as important disease-specific issues, will be considered in this chapter. Transition from paediatric to adult health services Effectiveness of transition planning A review of the effectiveness of transition planning has confirmed improved health outcomes when specific interventions to improve coordination between adult and paediatric services are implemented. Most research in this field has been undertaken with young people with diabetes, and many of the outcome measures relate to that condition. The principles of transition planning and the potential benefits are, however, likely to be generalisable to other LTCs that present in childhood. Young people with serious LTCs are among the most complex and high-risk patients to care for in adulthood, and it is important to work closely with them as they move to adult services, to try to improve their long-term outcome. General principles

of transition planning Paediatric services are organised and delivered in a very different way to adult medical services. They encompass a period of life that spans from infancy to independence, and progress from taking parents' views as paramount to needing to recognise Historically, childhood illnesses were characterised by a series of acute episodes, often infective, on a background of an otherwise healthy patient. Adult medicine traditionally comprised patients with progressive conditions, and increasing pathology with advancing age. A number of factors have led to the recognition that boundaries between adult medicine and paediatric care are not clear-cut, and recent evidence has confirmed that anticipating and carefully planning the transition of children with long-term conditions (LTCs) into adult services improve care and outcomes. About 14% of children in the developed world are diagnosed with an LTC and in the majority of patients the disorder will persist into adulthood. Common illnesses include asthma, epilepsy, congenital heart disease, diabetes and childhood cancer (Box 31.1). Similar trends are developing worldwide, with increasing survival rates Fig. 31.1 Reasons to consider transition planning. Transition planning Survivors of previously lethal conditions New clinical specialties, e.g. adult congenital heart disease High rates of loss to follow-up Ongoing medical problems, or complications of previous therapy Complex physiological and behavioural changes Adolescence associated with non-adherence Improve outcome Fig. 31.2 Lifestyle changes during transition to adulthood. Autonomous Dependent Transition Employment Education Graduation Financially independent Financially dependent Employment/ benefits Adult relationships, family planning Pre-pubertal Puberty Independent living Live with family Leave home Patient takes responsibility Parents oversee care Loss of parental control 31.1 Important long-term conditions of childhood that affect adult health Neurology • Epilepsy • Cerebral palsy • Duchenne muscular dystrophy Respiratory medicine • Cystic fibrosis Endocrinology • Diabetes mellitus Cardiology • Hypertrophic obstructive cardiomyopathy • Congenital heart disease Nephrology • Renal insufficiency • Renal transplant Gastroenterology • Inflammatory bowel disease Rheumatology • Inflammatory rheumatic disease • Osteogenesis imperfecta • Hypophosphataemic rickets Oncology • Survivors of childhood cancer Infectious disease • HIV/AIDS • Malaria

needs consideration in terms of prescribing and drug doses. The general advice is that when prescribing a dose per kilogram, the optimal weight for height rather than actual weight should be used for obese young people. A systematic approach to transition planning Several steps need to be undertaken to develop a successful programme for transition of care. The key components are summarised in Box 31.2. The first step is to establish a policy in consultation with young people and train staff in the policy. Subsequently, systems need to be developed to identify patients in need of transition and track them as they pass through the programme. Adult health-care providers need to be identified and processes developed for introducing the young person to the adult team. This should be followed by written communication between the paediatric and adult teams, and then a first consultation with the adult team at which the transfer can be reviewed and the wishes of the young person. After transition, young people move from medical services that have been family-centred and focused around maximising the child's development, to a service that encourages patient autonomy, in which employment and reproduction are important measures of outcome. At the same time as undergoing transition within medical services, young people are making multiple other transitions in their lives as they move from a dependent to an independent way of living (Fig. 31.2). They often move away from the family home, and parents

who formerly held responsibility for patient management, coordination of care, communication and consent to treatments will be demoted to an advisory role. Paediatric services are not well placed to meet this change in focus from the patient as a child to the patient as an independent adult, and young people benefit from the move to adult services as long as their specific needs as a young adult are recognised. Principles of prescribing during transition Hepatic drug metabolism increases from neonatal levels during childhood, eventually decreasing to adult levels after puberty. Once puberty has been completed, teenagers can be considered, in pharmacokinetic and pharmacodynamic terms, to behave like adults. It is important to remember that many young people have considerably lower body mass and therefore body mass index (BMI) than adults, and care needs to be taken to avoid excessive dosage in physically smaller patients. Likewise, obesity 31.2 Core elements in developing a transitional care programme Establishing transition policy • Develop policy, with input from young people • Train staff in operation of policy Tracking and monitoring • Establish process to identify patients • Develop systems to track individual progress • Incorporate transition planning into clinical care Transition readiness • Identify suitable adult care provider • Establish process for introduction to adult team • Provide written information about joint first consultation Transition planning • Ensure communication between paediatric and adult teams • Identify need for handover consultation • Prepare written medical handover: Diagnosis Current treatment Previous key issues • Send relevant information in advance • Provide information and community support Transfer to adult services • Arrange first consultation • Review transfer package with team • Identify concerns of young person • Review young person’s health priorities • Update medical summary and emergency care plans Integration into adult services • Communicate with paediatrics and confirm transfer • Help young adult to access other adult services • Continue individualised care plan tailored to young person • Seek feedback from young adult about transition 31.3 Key features in assessing readiness for transition to adult services (K) Knowledge

1. Describes condition, effects and prognosis
2. Understands medication purpose and effects
3. Understands treatment purposes and effects
4. Knows key team members and their roles (S) Self-advocacy
5. Can attend part/whole clinic appointment on their own
6. Knows how to make appointments/alter appointments
7. Has understanding of confidentiality
8. Orders repeat prescriptions
9. Takes some/complete responsibility for medication/other treatment
10. Knows where to get help (H) Health and lifestyle
11. Understands importance of diet/exercise/dental care
12. Understands impact of smoking/alcohol/substance use
13. Understands sexual health issues/pregnancy/sexually transmitted infections (A) Activities of daily living
14. Self-care/meal preparation
15. Independent travel/mobility
16. Trips/overnight stays away from home
17. Benefits/financial independence (V) Vocational
18. Current and future education/impact of condition on career plans
19. School attendance and performance
20. Work experience and access to careers advice

21. Outside activities and interests
22. Disclosure to school/employer (P) Psychosocial
23. Self-esteem/self-confidence
24. Body/self-image
25. Peer relationships/bullying
26. Support networks/family/disclosure to friends
27. Coping strategies (T) Transition
28. Understands concept of transition
29. Agrees transition plan
30. Attends transition clinic
31. Visits adult unit (if appropriate)
32. Sees primary care team/other clinical staff independently

1290 • ADOLESCENT AND TRANSITION MEDICINE follicle production, ovulation and menstruation, as described on page 652 and shown in Figure 18.14. Other hormonal changes in all adolescents include a rise in adrenal androgens and a rise in growth hormone, which in turn stimulates production of insulin-like growth factors 1 and 2 (IGF-1 and IGF-2). Insulin production also rises by about 30% during puberty. These hormonal changes contribute to the biological, morphological and psychological changes seen during the teenage years. Adolescence (as opposed to puberty) comprises not only the physical changes of puberty, but also the wider emotional and psychological changes of progression into early adulthood. The emotional and psychological changes are associated with physical maturation but also with sociocultural influences. The normal feelings and behavioural development of normal adolescence are complex but tend to follow fairly predictable patterns. Physical changes In girls, there is an increased rate of growth, followed soon after by the development of breasts and pubic hair. Menstruation typically starts after the rate of growth has peaked. In boys, puberty begins with testicular enlargement, followed soon after by a growth spurt and the development of pubic hair. In clinical practice, Tanner staging is used as a method of documenting progression of physical changes that occur during puberty (Fig. 31.5). The average age at onset of puberty in the UK is about 11 years in girls and 12 years in boys but normal puberty has a very wide range of onset. Factors that are important in predicting age of onset of normal puberty include family history (age of onset is strongly predicted by the parents' pattern of onset) and body mass, with heavier children entering puberty at a younger age. The current trends towards improved nutritional status and increased obesity in particular are driving earlier onset of puberty. Delayed puberty is defined to have occurred when the age at onset is more than 2.5 standard deviations above the national average, which in the UK is about 13 years in girls and 14 years in boys. If puberty is delayed beyond this point, investigations may be needed to determine the underlying cause, as detailed on page 653. Many children who have had long-term health conditions during childhood experience a delayed onset of puberty because chronic ill health slows longitudinal growth and causes functional hypogonadotrophic hypogonadism. Glucocorticoid therapy also contributes to growth retardation in children with chronic inflammatory diseases. An X-ray of the left wrist can be used to assess bone age accurately, and a bone age that is more than 2 years behind the chronological age should prompt consideration of further investigations (p. 654). Fig. 31.3 Timing of transition. Transfer to adult service with transfer package Prepare for adult model Discuss transfer Make aware of transition planning 12 years 14 years 16 years 18 years 20 years 22 years 24 years 26 years Integrate into adult care Initiate planning a care plan developed. There should subsequently be written communication between the adult and paediatric

teams to confirm that handover has occurred, followed eventually by integration of the young person's care into the adult service. A number of organisations have published guidelines to planning transition services. Two of the best known include the 'Ready Steady Go' programme in the UK and the American Academy of Paediatrics' 'Got Transition' (see 'Further information'). Details of the sorts of competencies that a young person might need before making a full transition to adult medical services are outlined in Box 31.3. When should transition happen? The optimum timing for transition is not specifically defined; it is a process that evolves over a number of years, during which puberty and then adolescence occur. Transition should generally be initiated at around 12 years of age. Completion time then varies from person to person, also depending on the model of adult services available. Most commonly, full transition occurs between 16 and 18 years of age (Fig. 31.3). This coincides with many other areas where young people are considered to have made the transition to adulthood, such as the completion of formal education. Marriage and children often follow. Functional anatomy and physiology Puberty and adolescence are developmental stages through which children progress during the second decade of life. During this phase, several physical, biochemical and emotional changes occur. The most important are discussed in more detail below. Endocrine changes The hormonal and physical stages of progression through puberty in males and females are summarised in Figure 31.4. Puberty is initiated by pulsatile increases in gonadotrophin-releasing hormone (GnRH) by the hypothalamus, which in turn stimulates pulsatile release of luteinising hormone (LH) and follicle-stimulating hormone (FSH) by the pituitary. In males, the increased production of LH stimulates Leydig cells in the testes to produce testosterone, and FSH acts on Sertoli cells to stimulate sperm production, as described on page 651 and shown in Figure 18.13. The rise in testosterone increases skeletal growth, promotes development of the male genital organs and stimulates growth of pubic, facial and axillary hair. In females, FSH and LH act on the ovary to promote

Fig. 31.4 Hormonal events of puberty. A In the ovary, FSH acts on granulosa cells to stimulate oestrogen production, whereas LH acts on theca cells to stimulate progesterone production. Androgens are also produced in small amounts by theca cells in response to LH (not shown). B In the male, LH acts on interstitial Leydig cells to stimulate testosterone production. FSH with testosterone acts on Sertoli cells to stimulate spermatogenesis. (ACTH = adrenocorticotrophic hormone; FSH = follicle-stimulating hormone; GnRH = gonadotrophin-releasing hormone; LH = luteinising hormone) From Smith RP. Netter's Obstetrics and gynecology, 2nd edn. Philadelphia: Saunders, Elsevier, Inc.; 2008. Acne appears Axillary hair appears Breasts develop Uterus enlarges Menstruation begins Pubic hair appears Vaginal epithelium cornifies Epiphyseal union hastened Body contours rounded Adrenal cortices ACTH GnRH LH and FSH Prolactin Pituitary LH and FSH secretion increased Higher cerebral centres 'trigger' puberty Reticular zone enlarges Adrenal androgens increased Oestrogen produced Progesterone increased Ovaries A Acne appears Facial hair appears Musculature develops Larynx enlarges (voice deepens) Axillary hair appears Some breast enlargement may occur Pubic hair appears Penis, prostate and seminal vesicles enlarge Epiphyseal union hastened Adrenal cortices ACTH GnRH LH and FSH Prolactin Pituitary LH and LSH secretion increased Higher cerebral centres 'trigger' puberty Reticular zone enlarges Adrenal androgens increased Testosterone increased Testes B

 Fig. 31.5 Tanner staging of puberty. Tanner stage I II III IV V Female Breast Pubic hair Pubic hair Mature stage. Projection of papilla with recession of areola to contour of breast Projection of areola and papilla to form mound above breast Further enlargement of breast and areola with no separation of contours Elevation of breast and papilla as a small mound Pre-adolescent Dark, coarse and curled hair extending to inner thighs Darker, coarse and curled hair but covering smaller area than in adult. No spread to medial surface of thighs Darker, coarse and curled hair Sparse, long and straight None Pre-adolescent None Penis, testes and scrotum of adult size Further growth in length and width of penis, testes and scrotum Growth of penis and further growth of testes and scrotum. Skin of scrotum becomes darker and more wrinkled Growth of testes and scrotum. Skin on scrotum reddens and becomes wrinkled Genitalia Male Dark, coarse and curled hair extending toward umbilicus Darker, coarse and curled hair but covering smaller area than in adult Darker, coarse and curled hair Sparse, long and straight Cognitive and behavioural changes As young people move from their early teenage years to later adolescence there is a move away from the family towards personal independence. This is often characterised by change from a self-centred focus, associated with a sense of awkwardness and worries about being normal, towards increased self-confidence and an awareness of weaknesses in parents and others in authority. In late adolescence, young people reach a stage of self-reliance, increased emotional stability and improved ability to think ideas through. Finally, young adults begin to develop firm belief systems, autonomy and independence. With time, there is reduced conflict with parents and other figures in authority and full maturity develops. In terms of cognition, there is a transition from being mostly interested in the present, in short-term outcomes and instant gratification, through to increased concern for the future and a greater focus on one’s longer-term role in life. Sexuality and relationships clarify during adolescence, and individuals move from early awkwardness and uncertainty to a firmer sense of their sexual identity, and then development of more serious and longer-term relationships. In terms of morals and values, young people move from a period of risk-taking behaviour and experimentation through to understanding the potential consequences of such behaviour for their future health and well-being. Young adults develop a greater capacity for setting personal goals and an increased focus on self-esteem. Finally, family, social and cultural traditions regain some of their previous importance, and by the time young people emerge from adolescence, they have usually developed insight and a greater focus on self-esteem and long-term well-being. It is the development of these more mature personality traits that are important for the more active role in health care that is needed to function well within an adult model of medicine. Some teenagers do vary slightly from these broad patterns but the feelings and behaviours described are, in general, considered normal for each stage of adolescence. Understanding these changes in emotional and psychological behaviour underpins the approaches that are needed to meet the challenges of managing long-term conditions in older teenagers and young adults.

assessment. Often, the first transition appointment is undertaken jointly with the paediatrician in a specialist transition clinic and this will enable a thorough face-to-face handover of all the key facts. A detailed transition referral letter should clearly describe the diagnosis, current and previous treatments, and key interventions that have been undertaken while the patient was under the care of paediatric services. It is important to check the main details with the young person and to make sure there are no other factors that they feel are of relevance. There are a few features in the

history that merit special attention, particularly at the first consultation, as outlined in Box 31.5. Many young people with an LTC attend their first adult outpatient department consultation with their parents, and it is important either to create a time to ask personal and lifestyle-related questions separately from the main history – that is, privately – or to make sure that these can be confidentially explored in future. Investigations Several changes take place during adolescence in terms of skeletal growth, organ development and body composition, which can influence the interpretation of results. Examples include fusion of the epiphyses as puberty progresses, increases in bone mineral content and density as the skeleton grows, and changes in the reference range of certain biochemical tests. Most of these changes occur gradually during puberty and there are rarely abrupt alterations in adult biochemical concentrations. It is important to use age-adjusted biochemical reference ranges until puberty has been completed. Several biochemical changes take place in the composition of body fluids between infancy and puberty. Some of the key changes in biochemical markers are outlined in Box 31.4. More detail on reference ranges for specific analytes is provided in Box 35.9 (p. 1363). The elevation in alkaline phosphatase (ALP) levels during adolescence relates to the bone isoenzyme produced by osteoblasts in the growing skeleton during the growth spurt. Further investigations for raised levels of ALP are not required in adolescence, as long as other liver function tests, including γ-glutamyl transferase (GGT), are normal. In general, under normal physiological conditions, the reference ranges of most biochemical tests remain fairly constant between puberty and menopause in women and between puberty and middle age in men. Clinical assessment The initial patient consultation at transition is of vital importance for establishing a potentially life-long professional relationship with the patient, as well as identifying key features in the history, examination and assessment of their overall needs. Parents commonly attend a first adult appointment with their son or daughter, and it is usually necessary to allow longer for this initial 31.4 Biochemical changes during transition Analyte Comment Alanine aminotransferase (ALT) Increased during adolescence Activity may continue to rise, at least in men, until middle age Alkaline phosphatase (ALP) Activity higher in infancy, decreases during childhood, and rises again with skeletal growth during puberty Peak in females at median 11 years and in males at 13 years Levels decrease rapidly after puberty, particularly in girls; adult levels achieved after epiphyses fused Insulin-like growth factor 1 Levels 30% higher during adolescence Serum creatinine Increases steadily from infancy to puberty parallel to development of skeletal muscle; until puberty, there is little difference in concentration between males and females Uric acid concentration Decreases from high levels at birth until 7–10 years of age, then increases, especially in boys, until 16 years 31.5 Features in transition assessment Features of history and/or examination Reason History* Main/presenting condition and detailed review of clinical course Understand detail and severity of illness; understand which treatments have been undertaken and which have been successful Family history: Draw family tree of all first-degree and any relevant/affected second-degree family members Many long-term conditions arising during childhood have significant genetic and familial factors to be taken into account Current drug therapy, significant previous therapies Many drugs have significant long-term implications, levels may need monitoring, may have teratogenic effects to consider Any surgical or other relevant medical history Understand which treatments have been undertaken and which have been successful Pubertal status/age of menarche Helps assess disease severity plus patterns of growth Informs about patient’s reproductive health and family-planning wishes Social history: In education or in work? Receiving appropriate benefits/support? Financial or other practical concerns? Living with parents/left home? Assesses wider effects of patient’s health on their independent living, as well as their financial and practical circumstances Can be a proxy measure of disease severity and

identifies their support mechanisms, which also helps in the assessment of their current needs Systems enquiry Any other related/unrelated symptoms or problems Physical examination Height, weight, calculation of body mass index Blood pressure Urinalysis if relevant Assessment of pubertal status General physical examination Although young people are often accompanied by a parent, examine them separately and use this opportunity to consider asking about private matters, such as partners, sexual activity, and drug or alcohol use *Throughout the first consultation, confidence and competence in decisionmaking/capacity to consent should be assessed. If there are concerns about capacity, clarify key decision-makers.

1294 • ADOLESCENT AND TRANSITION MEDICINE reported rates vary according to the method of assessment. Teenagers may also have varying adherence levels within their treatment regimen. An important example is in patients who have undergone organ transplantation, in whom low adherence to immunosuppressive medication is a significant cause of graft rejection and may cause death. Adherence merits careful consideration when caring for adolescents and young adults, and focusing on strategies to improve adherence at this initial stage of patient management can deliver life-long improvements in health outcomes. Young teenagers mainly believe in things that they have directly experienced and do not fully appreciate the unseen consequences of not taking their medications. In time, Presenting problems in transition medicine Problems with adherence Adherence is defined as ‘the extent to which a person’s behaviour, in terms of taking medications, following diets, or executing lifestyle changes, coincides with medical or health advice’. The term ‘adherence’ is used in preference to ‘compliance’ because it focuses on whether a person actively adheres to the regimen rather than passively follows the doctor’s orders. It also implies partnership and cooperation between the patient and the care-giver. More recently, clinicians have moved to seeking patients’ concordance with management plans. Concordance refers to a consultation process that has an underlying ethos of shared decision-making. It has become clear that current levels of adherence do not deliver the full benefits of medication. Historical paternalistic medical practice does not maximise the chances of patients adopting the changes and treatments they need to improve their outcomes. Reaching a concordant position with patients involves a range of approaches (such as patient-centredness or shared decision-making) and a number of specific actions (such as exploring anxieties about medication side-effects, individualising regimes to suit the patient’s lifestyle, offering a range of treatment options) and has not been evaluated comprehensively. Adherence to clinic attendance, investigation and treatment often falls significantly in adolescence and during transition to adult services. Measurement of adherence is challenging and 31.6 Factors affecting adherence Negative factors • Older adolescent • Mental health issues with care-giver • Family conflicts • Complex therapy • Medication with side-effects • Denial of illness Positive factors • Positive family functioning • Close friends • Internal locus of control • Treatment with immediate benefits • Patient’s belief in seriousness of illness and efficacy of treatment • Physician empathy Simplify regimen • Use once daily/twice daily regimes if possible • Match regimen to bedtime and meals • Use pill box or alarms on phone • Organise services around patient (combined clinics, flexible timing and appointments) Impart knowledge • Share decision-making • Provide clear instructions: Limit to three or four major points Use simple, everyday language Use written information or pamphlets and verbal education at all encounters • Supply addresses of quality websites • Provide advice on how to cope with medication costs Modify patient beliefs • Empower patients to self-manage their condition: Ask about their needs Ask what might help them become and remain adherent Ensure they understand the risks of not taking their medication Address fears and concerns about taking the medication Provide communication •

Improve interviewing skills • Practise active listening • Provide emotional support – treat the whole patient and not just the disease • Provide clear, direct and thorough information • Elicit the patient's input in treatment decisions • Allow adequate time for patients to ask questions • Build trust Leave the bias • Learn more about low health literacy and how it affects patient outcomes • Consider care of ethnically and socially diverse patient populations • Acknowledge biases in medical decision-making (intentional or unintentional) • Address dissonance of patient–provider race/ethnicity and language • Take extra time to overcome cultural barriers • Ask specifically about attitudes, beliefs and cultural norms around medication • Use culturally and linguistically appropriate targeted patient interventions • Increase engagement, activation and empowerment • Tailor education to the patient's level of understanding Evaluate adherence • Direct: Number of repeat prescriptions Biomarkers of response Measurement of drug levels • Indirect: Self-reporting: 'When did you last forget your medicine?'; 'How often have you forgotten your medicines this week?' 31.7 SIMPLE strategies to improve adherence Excerpted with permission from the American College of Preventive Medicine. Medication Adherence: Improving Health Outcomes Time Tool: A Resource from the American College of Preventive Medicine. 2011. Retrieved from https://webmail2.tst.nhs.uk/go/www.acpm.org/?MedAdhereTTProviders.

are at greater risk of undertaking harmful behaviour and there is evidence that a poor long-term health outlook is associated with risk-taking behaviour earlier in life. Globally, the leading causes of death among adolescents are road injury, human immunodeficiency virus (HIV) infection, suicide, lower respiratory infections and interpersonal violence; many of these deaths are linked to risk-taking behaviours such as excess alcohol and drug intake. Quite apart from mortality, there are other significant adverse events linked to risk-taking behaviour in adolescents: excess alcohol ingestion is also associated with non-fatal road traffic accidents, unwanted and unprotected sexual activity, and violence as both perpetrator and victim. There are a number of theories about the neurodevelopmental changes associated with these behaviour changes. At around 11 years of age, the prefrontal cortex (PFC) and parietal lobes begin a period of pruning of neuronal axons. It is theorised that these changes represent the start of the process of increasing frontal lobe control. A separate process that occurs at the same time predisposes the adolescent to risk-taking behaviour and impulsivity: frontostriatal reward circuits mature relatively early and encourage the adolescent towards adult activities such as alcohol and drug use, and sexual intercourse, which carry potential health risks. At this stage, the PFC has not yet matured to the point where the individual can assess risk adequately. The PFC and its connections are structurally unable to provide sufficient control. It is thought that this maturational gap in PFC control of the pleasure-seeking brain systems is responsible for the risk-taking lifestyle that characterises the period of adolescence. A number of studies have investigated personality and other factors that contribute to different risk-taking behaviours during adolescence: essentially, younger adolescents and females tend to rate activities as being more risky, and are therefore less likely to undertake them. Older males and those of lower educational status are higher risk takers. Specific protective factors include high self-esteem and a strong orientation to an internal locus of control; young people who feel they have less control and influence over themselves and their behaviour are more likely to undertake high-risk activities. Many teenagers with serious long-term health conditions are disempowered in a number of ways and there is evidence that they are predisposed to be high risk takers during adolescence. Examples of risk-taking behaviour include not only alcohol and drug intake, but also

non-adherence to medicines and other aspects of health care, such as diet in diabetes. It is not easy to affect behaviour during this period of nonadherence. Isolated educational intervention is not sufficient to improve outcome; for example, there is a wealth of evidence showing that teenagers know about the behaviours needed to prevent transmission of HIV, but many do not adhere to this advice. Long-term health-care providers have an invaluable role in supporting adolescents during this period of their development as adults, as many important health-related and lifestyle habits are established during this period: more than 90% of smokers start smoking in adolescence, and life-long habits around eating and exercise are laid down during the teenage years. Focusing on the needs of the emerging adult for autonomy and using the highest levels of communication and patient engagement significantly improve outcomes for patients with LTCs. Unplanned pregnancy In many parts of the world, females commonly undergo their first pregnancy during or just after adolescence. The median age for 31.8 History-taking in adolescent patients: risk-taking behaviours ('HEADS') Home life • Relationships • Social support • Household chores Education • School • Exams • Work experience • Career • University • Financial issues Activities • Peers, people that patients can rely on • Exercise and sport Driving • Aged 16 if disabled Drugs • Cigarettes and alcohol: how much, how often • Non-prescription drugs Diet • Nutritional content (calcium, vitamin D) • Weight • Caffeine (diet drinks) • Binges/vomiting Sex • Concerns • Periods • Contraception (in relation to medication) Sleep • Amount • Difficulty getting to sleep • Frequent waking • Early waking? Suicide • Depression • Mood • Disabled adolescent men high-risk From Segal TY. Adolescence: what the cystic fibrosis team needs to know. J R Soc Med 2008; 101(Suppl 1):15–27. adolescents learn to develop hypothetical thinking and to analyse more complex information and decision-making. The ability to engage in formal thinking is inconsistent at first, and at times of stress (such as during an illness) adolescents may regress to more simple ways of problem-solving. Despite their maturing skills, they may remain self-centred and feel invincible. Factors that positively and negatively affect adherence are outlined in Box 31.6. Interventions to improve adherence are summarised in Box 31.7. Recent literature suggests that two-way communication between patients and professionals about medicines leads to improved satisfaction with care, knowledge of the condition and treatment, adherence, health outcomes and fewer medicationrelated problems. Younger adults and those coming to adult services following transition from paediatric services have very different expectations in terms of the nature of the patient–doctor relationship and are more likely to require a more collaborative approach to development of management plans to maximise their concordance with treatment in the long term. High-risk behaviour The high-risk behaviour that can be undertaken by adolescents is well documented and is seen across many cultures. It is important to assess this by history-taking at the time of transition (Box 31.8) Adolescents who have had LTCs during childhood

1296 • ADOLESCENT AND TRANSITION MEDICINE controlled with first-line anticonvulsants in around 80% of cases. Young people who still have epilepsy or are on anticonvulsant therapy as they progress into adulthood are more likely to have underlying structural brain disease, such as cerebral palsy, or have more complex or syndromic epilepsy. In many of them, epilepsy may be associated with learning difficulties or other neurological conditions. Epilepsy presents several problems during transition. Adherence to medication can be an issue and patients with low adherence to epilepsy medicines have higher mortality, higher hospital admission rates and higher emergency department attendances. Conversely, high adherence rates at initiation of epilepsy therapy are associated with improved long-term seizure freedom and higher seizure freedom at 4 years. Epilepsy can also affect employment options for young people, as about 30% of patients still

have breakthrough seizures while on treatment. Certain types of employment, such as working within the emergency services or armed forces, or becoming a pilot or driver of a heavy goods vehicle, may therefore not be possible. Driving restrictions may also limit options for some other occupations (p. 1103). Young women with epilepsy should be advised that oral contraceptives are less effective with enzyme-inducing antiepileptic drugs; they should also be made aware of the risk of teratogenicity with many antiepileptic drugs, most notably sodium valproate, which should be avoided in pregnancy if at all possible. Pre-conceptual counselling is desirable for all young girls with epilepsy and pre-conceptual folic acid supplementation is advisable to reduce the risk of neural tube defects. Alcohol use in moderation does not affect seizure control in the majority of patients, but withdrawal from alcohol in dependent patients is epileptogenic and heavy alcohol use should be discouraged. Information about marijuana and epilepsy risk is lacking, but regular marijuana use and excess drinking are associated with poor adherence to medication regimes and increased seizure risk. Cerebral palsy Cerebral palsy comprises a range of non-progressive neurological impairments, present from the time of birth or arising in early childhood. Although the neuropathology is non-progressive, the manifestation of problems can evolve, with progressive motor dysfunction related to increased spasticity and possibly progressive seizure activity. Patients with severe cerebral palsy present specific problems during transition and early adulthood. They may be paraplegic or quadriplegic and most non-ambulatory individuals have significant intellectual disability. This group of patients will be unable to live independently during adulthood and need ongoing long-term care. Delivering medical care to these individuals poses several problems, including practical issues such as consideration of capacity and consent to treatment. Other comorbidities include gastro-oesophageal reflux (often related to abnormal lower oesophageal function), seizures, and feeding difficulties often requiring gastrostomy. These individuals usually require a complex care package involving many members of the multidisciplinary team. There are also risks of abuse and neglect in the care of adults with severe disability and this needs to be borne in mind when considering atypical problems or unusual presentations in this vulnerable patient group. Depending on the severity of the patient’s condition, end-of-life care may need to be discussed and planned with the family and other care-givers. 31.9 Adolescent pregnancy rates Region Pregnancy rate (/1000 women aged 15–19 years) Tanzania

Kenya

Jamaica

Mexico

South Africa

USA

Pakistan

New Zealand

UK

Spain

France

Japan 5.3 Italy 4.5 South Korea 2.2 first pregnancy varies from 19 years in India and parts of Asia, through to 25 years in the USA and around 30 years in Australia and Western Europe. Teenage pregnancy rates are high across the world (Box 31.9). Information from the UK suggests that 1 in 6 pregnancies is unplanned, and 1.5% of women between the ages of 18 and 45 face an unplanned pregnancy each year. It is therefore vital to anticipate and discuss the issues surrounding reproductive health with all young people before and during transition, as well as during early adulthood. Young people with serious LTCs have a number of additional factors to be taken into consideration when discussing reproduction, and these discussions need to take place long before a family is planned. General physicians do not need to be able to undertake complex genetic counselling and investigation, but should be able to provide advice about the recurrence risk of common inherited conditions, as well as that of the more common multifactorial LTCs, many of which have an inherited or genetic component. Clinical presentations In almost every clinical setting, there is the potential for a young adult with a serious LTC that has arisen during childhood to present to adult physicians. Medical services can improve the care and outcome for this vulnerable group by planning a systematic approach to transition, as described above, and by focusing the clinical consultation on issues of relevance and importance to each particular patient. The key issues to consider for a number of the most common LTCs of childhood are discussed below. Neurological disease Epilepsy Epilepsy is a chronic disorder, the hallmark of which is recurrent, unprovoked seizures (p. 1097). Epilepsy that has presented during childhood, as opposed to adulthood, is less often associated with underlying central nervous system malignancy and is well

Muscular dystrophy The muscular dystrophies are a group of diseases that cause progressive weakness and loss of muscle mass. The disorders differ in terms of which muscle groups are affected, the degree of weakness, and the rate of disease and symptom progression. All the inherited muscular dystrophies that present during childhood need active management during transition (p. 1143). One of the most important and most severe conditions is Duchenne muscular dystrophy (DMD), which is associated with a progressive decline in mobility, coupled with cardiac dysfunction due to cardiomyopathy, and respiratory failure requiring respiratory support as the disease progresses. Physicians should ensure that the whole family are aware of the wider genetic issues. Female carriers of the DMD gene can suffer muscle fatigue and are at risk of cardiomyopathy, as well as there being obvious risks for their male offspring. On average, patients with DMD survive until their late teens to early twenties, and those with less severe muscular dystrophies, such as Becker dystrophy, survive until their thirties. Although fertility is reduced, young men with these conditions may themselves father children. Male offspring will be unaffected but all female infants of affected males will be carriers. As the muscular dystrophies progress, a complex package of care involving a multidisciplinary team is necessary; it should include respiratory input to assess the need for ventilatory support, which is a common endpoint for many patients. At the present time, there is no definitive treatment. Glucocorticoids (0.75 mg/ kg/day) have been shown to improve muscle strength and are frequently used, but carry an increased risk of osteoporosis and vertebral fractures. New therapeutic approaches are being developed with the aim of ameliorating disease progression in patients with nonsense mutations (p. 42). One involves

the use of drugs such as ataluren, which promotes binding of transfer RNA (tRNA) molecules at the site of stop codons with a mismatch in one base (near-cognate tRNAs). These cause a full-length protein to be produced with an amino acid substitution rather than a truncated non-functional protein. Patients with DMD usually require social and financial support. It is important to consider end-of-life care plans with patients and family members. Involvement from palliative care teams, as well as psychological, spiritual and wider non-medical support, is essential. Respiratory disease Cystic fibrosis Cystic fibrosis (CF) is a single-gene autosomal recessive disorder that affects about 1 in 2000 to 1 in 3000 individuals of Caucasian descent (p. 580). Clinical manifestations are caused by defects in an ion transporter termed the cystic fibrosis transmembrane conductor regulator (CTFR) protein. With improved supportive care, the median survival in the UK is now more than 50 years. It is well recognised that young people with life-limiting conditions face particular challenges during transition: individuals often exhibit high-risk behaviour during adolescence, and this was particularly true in the past when long-term survival rates were poor. The rates of non-concordance with medication and with time-consuming physiotherapy and nebuliser regimes are high and adversely affect outcome and survival. Exercise tolerance and employment are likely to be restricted as time progresses, and patients need particular support managing the slow decline in function and well-being that occurs throughout their adult lives. In terms of fertility and child-bearing potential, the picture is complex and merits detailed discussion with patients. It is not often discussed openly by the paediatrician or during childhood, other than with the parents when the patient is young. The vas deferens is absent in 98% of males with CF and seminal vesicular dysfunction means that ejaculates are low in volume. While boys are infertile, newer reproductive therapies, such as the availability of intracytoplasmic sperm injection, mean that fatherhood is possible. The opportunity for assisted reproduction should be discussed early so that people can make informed choices at an appropriate stage of their lives. Females with CF who have good nutritional status and reasonable health status have normal fertility and genetic counselling should be offered early. Contraception needs to be discussed with women who are not planning a pregnancy, since pulmonary hypertension is an absolute contraindication for the oral contraceptive pill (OCP). Women also need to be advised about the effects of antibiotics on OCP effectiveness. New orally available small-molecule therapies, including lumacaftor and ivacaftor, have recently been licensed; they can partially rectify functional defects in the CTFR and have improved outcome. These drugs are having a positive effect on symptom control and are potentially disease-modifying. Other therapeutic approaches, including gene therapy and mRNA editing therapies, are also being explored as treatments for CF. Common issues encountered during transition of CF patients are summarised in Box 17.34 (p. 581). Cardiovascular disease Congenital heart disease Congenital heart disease (CHD) is the most common congenital anomaly, affecting about 1% of live births. Among birth defects, CHD is the leading cause of mortality. Maternal illnesses such as rubella and injection of teratogenic agents during pregnancy, along with paternal age, all play roles in pathogenesis. Although some chromosomal anomalies, such as trisomy 13, 18 and 21 and monosomy X (Turner’s syndrome), are strongly associated with CHD, these account for only 5% of cases. Microdeletion and single-gene mutations can also be important, such as in DiGeorge syndrome (22q11.2 microdeletion). Overall, the most common congenital valvular anomalies are aortic and pulmonary stenosis. The most common structural anomaly is ventricular septal defect. There is a wide range of severity of CHD but many patients with life-limiting conditions (usually complex structural anomalies such as tetralogy of Fallot or hypoplastic left heart syndrome) survive to adulthood. Genetic counselling of affected individuals is important, as there is a 1–2% recurrence risk of any cardiac anomaly in offspring. Affected patients should be transitioned to a

cardiologist with experience in CHD since this has become a subspecialty in own right. More details are provided on page 531 and in Box 16.103 (p. 537). Hypertrophic obstructive cardiomyopathy Hypertrophic obstructive cardiomyopathy (HOCM, p. 539) is a genetic cardiovascular disease characterised by left ventricular wall hypertrophy, impaired diastolic filling and abnormalities of the mitral valve. These features can cause dynamic obstruction of the left ventricular outflow tract, diastolic dysfunction, myocardial dysfunction and an increased risk of supraventricular and ventricular tachyarrhythmias. HOCM is caused by mutations

1298 • ADOLESCENT AND TRANSITION MEDICINE necrosis, and angina or myocardial infarction arising from vasoocclusion or vasospasm. In addition to physical effects, children who have faced lifethreatening illness in childhood may experience psychological and family difficulties during adulthood. Cognitive impairment is more common in children who have received chemotherapy or radiotherapy to the brain, and problems can include lower IQ, problems with memory and attention, poor hand–eye coordination and behaviour/personality problems, combined with the wellrecognised and physical complications of cancer treatment in childhood. Increasing recognition of these issues has resulted in active monitoring programmes for survivors of childhood cancer, who are best seen in specialist ‘late effects’ multidisciplinary clinics, where teams include oncologists, psychologists and specialists from other relevant disciplines. Renal disease Chronic kidney disease (CKD, p. 415) accounts for some of the most complex long-term illnesses in childhood. The most common causes during childhood and adolescence are shown in Box 31.10. The primary pathology can be varied and many conditions have no specific treatment, but the overall approach to management of progressive renal insufficiency is the same. Internationally agreed definitions of CKD staging in children differ from those in adults and are summarised in Box 31.11. In adults the rate of albumin excretion is included in CKD definitions, as outcome correlates with the level of albuminuria, but in children similar data are lacking and so the staging system is based on glomerular filtration rate alone. The majority of children with affecting the genes that encode cardiac sarcomere proteins and is most frequently transmitted as an autosomal dominant trait. It may present for the first time during adolescence with cardiac arrest or sudden cardiac death. Predictive genetic testing is possible but challenging because of the large number of causal mutations. In clinical practice, careful analysis of the family history can be useful in identifying those at risk of inheriting the disease. If no gene anomaly has been identified within a family, first-degree relatives may need screening by electrocardiography (ECG) and echocardiography. Identification of a genetic anomaly is most helpful in allowing identification of family members who do not need echocardiograms or clinical follow-up. Children of affected parents should be screened every 3 years until puberty, and then annually until 20 years of age. If there is no evidence of HOCM in early adulthood, it is unlikely that the condition will develop in later life. Oncology Around 1 child in 500 will develop cancer by the age of 14 years. Leukaemia is the most common, accounting for about 33% of cases; central nervous system tumours are the next most common, accounting for around 25% of all childhood cancers. Fifty years ago, 75% of children diagnosed with cancer died, but overall survival rates now range from 75% to 80%. Between 60% and 70% of young adults who have survived childhood cancer will develop at least one medical disability, most commonly as a result of their therapy rather than their primary cancer. There is a 3–6-fold increased risk of a second cancer, with an absolute risk of about 10% before 50 years of age. It is therefore important for these individuals to be kept under surveillance during transition and beyond. Endocrine and reproductive disturbances are the most common late effects, affecting 40–60% of survivors. Infertility can be an issue in both males and females receiving cytotoxic

medications, unless it has been possible to store semen and ovarian tissue in advance of treatment. Other long-term risks include hypopituitarism, growth hormone deficiency and pubertal delay (especially in boys) from brain irradiation. Radiotherapy to the neck can cause hypothyroidism and increases the risk of thyroid cancer. Total-body irradiation offered as conditioning for bone marrow transplantation affects both ovarian and testicular function, and many of the chemotherapeutic agents used have adverse effects on fertility. Chemotherapy-induced ovarian failure is typically associated with high-dose alkylating agents such as cyclophosphamide, and this is an independent risk factor for premature ovarian failure. In recent years, patients have been offered ovarian and testicular tissue retention and fertility issues are being discussed with families during childhood, but often the patients themselves have limited levels of knowledge of the details. Chemotherapy and radiotherapy in childhood significantly reduce ovarian reserves. When combined with the progressive ovarian decline that occurs in all women throughout adulthood there is a significant risk of premature menopause or ovarian failure, with 8% of survivors affected. Young women need to be aware of these risks during their early adulthood to help with family and lifestyle planning; for example, they may wish to plan to have children earlier in their adult life rather than risking ovarian decline. Cardiomyopathy is another complication of anthracyclines such as doxorubicin and daunorubicin. Serious cardiac complications include arrhythmias, dilated cardiomyopathy from myocardial 31.10 Causes of renal impairment in childhood and adolescence • Obstructive uropathy • Renal hypoplasia/dysplasia • Reflux nephropathy • Focal segmental glomerular sclerosis • Polycystic kidney disease 31.11 Staging of chronic kidney disease (CKD) in children over 2 years of age* Stage Glomerular filtration rate (GFR) (mL/min/1.73 m2) Description

> “ 90 Normal or high

60–89 Mildly decreased 3a 45–59 Mildly to moderately decreased 3b 30–44 Moderately to severely decreased

15–29 Severely decreased

< 15 Kidney failure *Kidney Disease: Improving Global Outcomes (KDIGO) 2012 classification. Chronic kidney disease is defined either as GFR < 60 mL/min/1.73 m2 for > 3 months, regardless of whether other CKD markers are present, or as GFR < 60 mL/ min/1.73 m2 accompanied by evidence of structural damage or other markers of kidney abnormalities, including proteinuria, albuminuria, and pathological abnormalities in histology or imaging. In adults, albumin excretion is also included in CKD staging as the level of albuminuria correlates to outcome. These data are lacking in children and so albuminuria is not used to classify paediatric CKD.

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Diabetes Adherence and concordance with medication are a particular challenge in adolescents who have developed diabetes during childhood (p. 753). Studies of adolescents with type 1 diabetes have revealed that 25% were neglecting insulin injections, 81% were not following their diet, and 29% were not measuring their glucose level and were completing a daily diary with

fictitious results. Research has also shown that the outcome of diabetes is improved with a formal transition programme (Box 31.12). Good control of diabetes is particularly important during this phase since microvascular disease often emerges around time of transition, although it can occur sooner in patients with early-onset diabetes. CKD stage 3 or higher will ultimately require renal replacement therapy but the timescale for reaching this stage can vary widely. The mainstay of support is to delay progression by treatment of secondary factors that are known to be associated with progressive decline in renal function: namely, hypertension, proteinuria and anaemia. Organ transplantation Children requiring renal replacement therapy are the most common recipients of kidney transplants in childhood. Liver and heart transplantation, followed by lung and small bowel transplantation, are well-recognised but less commonly undertaken procedures. Non-adherence to immunosuppressive regimes during adolescence is a well-known risk factor for graft failure. The reported incidence of graft failure due to non-adherence is 10–15% but this is likely to be an under-estimate. Rates of non-adherence are highest among adolescents and young adults. As well as non-adherence to immunosuppression, non-adherence to testing and clinic attendance adversely affects the care and outcome of around 1 in 8 kidney transplant patients. Poor adherence is associated with patients with worse psychological status and family dysfunction; adherence has been shown to improve with education and increased motivational factors, as might be expected. This offers the opportunity to improve graft survival. At present, 50% of cadaveric grafts and around 68% of live donor grafts are still functioning 10 years post-transplant. There is no clear difference between children and adults in survival of transplanted kidneys. Many medications used in transplant medicine and in renal disease can have long-term effects on health. Inhibition of linear growth is seen even with low doses of glucocorticoids, such as 0.125 mg/kg/day on a long-term basis. Alternate-day regimes are generally considered preferable in childhood. The height reduction associated with long-term glucocorticoid use in childhood is dose-dependent, and even for children with asthma treated with inhaled glucocorticoids, an average height reduction of 1.2 cm is reported. This is can also be associated with delayed puberty, as well as an increased risk of osteoporosis in adulthood. Post-transplant lymphoproliferative disorders (PTLDs) are a well-recognised and potentially life-threatening complication in solid organ recipients. PTLD is the most common malignancy complicating solid organ transplantation, accounting for 20% of all cancers. They represent a range of lymphoproliferative disorders, from infectious mononucleosis and lymphoid hyperplasia to malignant lymphoma. Most cases of PTLD are associated with Epstein–Barr virus (EBV), leading to uncontrolled B-cell proliferation and tumour formation. Up to 10% of solid organ transplant recipients develop PTLD but the risk is almost four times higher in patients under 20 years of age, as opposed to those aged 20–50. This increased risk relates mainly to the development of EBV infection after transplantation; most adults are already EBV-seropositive at the time of transplantation and therefore at lower risk of this complication. The type of organ transplant that has been undertaken predicts PTLD risk, with the cumulative incidence over 5 years ranging from 1–2% in haematopoietic cell transplant and liver transplants, 1–3% in renal transplants, 2–6% in heart transplants and 2–9% in lung transplants to as high as 11–33% in intestinal or multi-organ transplants. The different rates possibly relate to the varying degrees of immunosuppression required. The incidence of PTLD is highest in the first year after transplantation, when it is associated with the highest levels of immunosuppression. 31.12 Impact of transition planning on outcome in diabetes Intervention Outcome measures Disease-specific education Lower HbA1C Generic education/skills training Fewer acute complications: Hypoglycaemia Admissions with ketoacidosis Transition coordinator Lower rate of loss to follow-up Joint paediatric/adult clinic Fewer chronic complications: Hypertension Nephropathy Retinopathy

Separate young adult clinic Improved self-management Improved disease-specific knowledge Out-of-hours phone support Improved screening for complications Enhanced follow-up Better quality-of-life scores Gastrointestinal disease Inflammatory bowel disease The prevalence of inflammatory bowel disease (IBD, p. 813) in childhood is increasing, and the incidence of Crohn's disease (CD) in particular is rising in both children and adults, probably due to currently undefined environmental factors. Current treatment aims are outlined in Box 31.13. Standard measures include exclusive enteral nutrition for 6–8 weeks using a whole-protein (polymeric) formula, which induces initial remission in 80% of children. This is equivalent to glucocorticoid therapy but offers improved nutritional status and superior mucosal healing. Glucocorticoids can also be used to induce remission, as well as to treat exacerbations, but should be followed up by immunosuppressive therapy with azathioprine or methotrexate. Adolescents and children are more likely than adults to require biologics and around 20% need treatment with tumour necrosis factor alpha (TNF-α) inhibitors such as infliximab or adalimumab. Around 20% of children with CD require surgery within 5 years of diagnosis; limited resections and stricturoplasty are considered best practice to preserve gut length and prevent short bowel syndrome (p. 707). Children with ulcerative colitis (UC) are more likely than adults to present with pancolitis (approximately 80% versus 40–50% in adults). Mild disease should be treated initially with oral 5-ASA preparations such as mesalamine or sulfasalazine. If the response is inadequate, oral glucocorticoids can be used, but caution must be exercised because of the adverse effects on skeletal growth and bone mineral density. Thiopurines such as 6-mercaptopurine or azathioprine are frequently used as steroid-sparing agents,

 and in young adults. Intravenous bisphosphonates are widely used in the treatment of children with osteogenesis imperfecta (those with long-bone deformities, vertebral compression fractures, and three or more fractures per year, in whom the benefit:risk ratio is thought to be positive), although the evidence base for prevention of fractures is poor and based on observational studies. There is much debate about whether continuing bisphosphonate therapy into adulthood is beneficial due to concerns about suppression of bone turnover in the long term. Affected individuals and their parents can find this change in treatment strategy confusing and it is important to explain the underlying rationale in order to manage expectations. Hypophosphataemic rickets Hypophosphataemic rickets is described in more detail on page 1052. Adherence to phosphate supplements and, to a lesser extent, vitamin D metabolites represents an important issue in optimising management during childhood and this becomes even more challenging in transitioning patients. While skeletal deformity does not progress following closure of the epiphyses, different problems arise in adolescent patients when there is suboptimal control of hypophosphataemia, including painful pseudofractures and arthralgia associated with enthesopathy. The renal phosphate leak tends to improve to an extent during adolescence and the requirement for phosphate is reduced, but most patients still require treatment with active vitamin D metabolites. Summary Young people who have suffered long-term conditions during childhood represent a particularly high-risk group of patients as they progress through adolescence to become young and, finally, mature adults. They bring with them specific medical risks and complications related to their previous medical treatment, and knowledge of these is important to identify the long-term complications of the therapies to which they have been exposed. They are a patient group that can display complex and often abnormal illness behaviour. Understanding this and implementing an effective process for transition from paediatric to adult services can reduce the significant risks that these patients face in early adulthood. As they mature and develop more adult intellectual and emotional behaviour patterns, the risks to their health and well-being reduce. Patients in transition can be a particularly challenging group to manage, but

investment of time and effort at this stage of their lives can be extremely rewarding and can bring significant improvements in long-term health-related outcomes. Further information Books and journal articles Atreja A, Bellam N, Levy SR. Strategies to enhance patient adherence: Making it simple. MedGenMed 2005; 7:4. Crowley R, Wolfe I, Lock K, McKee M. Improving the transition between paediatric and adult healthcare: a systematic review. Arch Dis Child 2011; 96:548–553. Segal TY. Adolescence: what the cystic fibrosis team needs to know. J R Soc Med 2008; 101(Suppl 1):15–27. Websites acpm.org/?Adherence American College of Preventive Medicine: detailed review of adherence. GotTransition.org Sample clinical tools and measurement resources for quality improvement purposes. uhs.nhs.uk More clinical tools and measurement resources. with progression to anti-TNF-α therapy for those who still do not respond. There is less evidence for efficacy of TNF-α inhibitors in adolescents with UC than those with CD; they seem to be effective at inducing an initial response but less useful for maintaining long-term remission, since a significant proportion of patients still require colectomy (20% at 1 year) or long-term glucocorticoids. Ciclosporin is probably more effective than infliximab in teenagers with refractory UC. As with other adolescents who have long-term conditions, adherence to medication is particularly important to reduce the risk of relapse. In terms of lifestyle advice, smoking is a particular risk since it increases both the rate and severity of relapses. Body image can be a particular challenge for young adults with IBD, and those with colostomies or fistulae, for example, can find this part of their illness particularly difficult. Delayed puberty and short stature are important comorbidities, partly related to medication side effects and also to the nature of the inflammatory bowel disease itself. Rheumatology and bone disease Juvenile idiopathic arthritis Juvenile idiopathic arthritis (JIA) is the term used to describe a wide variety of inflammatory rheumatic diseases that present during childhood (p. 1026). Oligoarticular juvenile arthritis has a good prognosis and often remits during adulthood, and so transitioning patients to adult rheumatology services may not always be required. The same does not hold true for systemic JIA and polyarticular JIA, which often require long-term immunosuppressive therapy through transition and beyond into adulthood. Smoking is a risk because it increases the activity of inflammatory disease and reduces the effectiveness of biologics. Adherence to and concordance with medication remain a challenge, as in other chronic diseases. Functional limitation secondary to joint damage may limit employment opportunities. Contraceptive advice is important in patients on methotrexate. Glucocorticoid-induced osteoporosis Osteoporosis is a complication of long-term glucocorticoid therapy that may be required in patients with inflammatory disease, transplantation and DMD. There is a paucity of evidence about best practice in glucocorticoid-induced osteoporosis in children and adolescents, but in general the teenage years are a period of considerable bone mineral deposition and offer a chance to enhance bone mineral density significantly. It is important to ensure adequate calcium and vitamin D intake and to supplement if necessary. Therapy with bisphosphonates can be considered in symptomatic patients, although the evidence base for prevention of fractures is poor. Osteogenesis imperfecta Osteogenesis imperfecta (p. 1055) typically presents with multiple low-trauma fractures during infancy and childhood. Although fractures become less common during adolescence due to the increase in bone mass, they still occur frequently during transition 31.13 Treatment strategy in Crohn’s disease • Induce and maintain clinical remission • Optimise nutrition • Optimise bone health • Optimise growth and pubertal progress • Minimise adverse drug effects

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Revision #1
Created 2026-01-08 16:24:59 UTC by Omar Ayman
Updated 2026-01-08 16:24:59 UTC by Omar Ayman