



Adolescence: what the cystic fibrosis team needs to know

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Adolescence can be defined as a period of biopsychosocial transition from a dependent child to an autonomous/independent adult, usually taking place between 13 and 20 years of age. There is considerable variation between individuals and cultures – in some it is truncated to circumcision. As a group adolescents make up 13% of the general population, but they probably account for about 30–40% of both the paediatrician and adult physician's workload, especially when dealing with a chronic disease of limited life span. Adolescence constitutes a large proportion of the life of a patient with cystic fibrosis (CF).

The treatment of adolescents poses specific challenges for treating disease and promoting health. In working with adolescents, the treatments for disease are played out against a background of rapid physical, psychological and social developmental changes; changes that produce unique communication and management challenges. At no other time of life are the physical and psychosocial elements of illness and behaviour so inextricably intertwined as in adolescence. This can make working with adolescents difficult; however, given the right skills – which can be learned! – practising medicine with young people can be extremely rewarding and productive.

The reasons for a distinct approach to medicine with adolescents

The doctor–patient relationship

Adolescence is the only time in paediatric medicine that the primary relationship is between the doctor and his patient, although this relationship is still within the context of the family.

Health behaviours are laid down in adolescence and continue into adult life

One of the most compelling arguments for a focus on adolescent health is that adolescence is a time when new health behaviours are laid down,

behaviours that will track into adulthood and influence health and morbidity throughout life. Health behaviours in childhood are dominated by parental instruction and shared family values. During adolescence young people begin to explore alternative or 'adult' health behaviours, including smoking, drinking, drug use, violence and sexual intimacy. The continuities between adolescent initiation of health behaviours and adult behaviour are well documented. For example, regular smoking rates rise from 1% at 11 years to 24% at 15 years, and over 90% of adult smokers began smoking during their teenage years. Equally importantly, health behaviours concerning exercise and food are laid down in adolescence and track into adult life.

Adolescents have unique needs in the management of health and illness

Dynamic and continued development in every aspect of a young person's life during adolescence means that young people have distinct needs in the management of illness and health. In clinical interactions with younger children, management decisions are made 'adult to adult' by health professionals in consultation with parents, and day-to-day disease management is generally undertaken directly by parents. When working with adolescents, the wishes, desires, knowledge base, capabilities and rights of the young person involved must also be taken into account – as must the fact that these are constantly evolving and changing! Different approaches are required to all aspects of the doctor–patient relationship. Examination of adolescents requires consideration of privacy and personal integrity as well as additional skills such as pubertal assessment. The effective treatment of illness in adolescence requires adept management of the issues regarding adherence, consent and confidentiality, and relationships between the young people and their family.

Table 1**Developmental changes occurring in adolescence. Amended table from *Nelson's Paediatrics*³³**

<i>Biological development (pubertal changes)</i>	<i>Psychological development</i>	<i>Social development</i>
<ul style="list-style-type: none"> • Breasts, testes and penis enlarges • Pubic and axillary and body hair • Menarche • Growth spurt (breast stage 2–3 age 11–12yrs, testicular volume 10 mL age 13–14) • Fat deposition • Secondary effects • Boys androgenic effects of muscle bulk • Body shape changes • Voice breaks • Accretion of bone mineral density • Acne • Blushing • Need for more sleep • Maturation of other organs (e.g. liver) 	<ul style="list-style-type: none"> • Thinking changes from concrete to abstract (logical) • See selves as 'bullet proof' • Sexual identity develops • Experimentation • May have homosexual peer interest • Reassess body image in face of changes • Growing educational abilities; develop vocational ideas • Learn to differentiate law and morality • Learn to control impulses • May develop fervent views 	<ul style="list-style-type: none"> • Realization of differences from parents → emotional separation • Peer identification • Exploratory behaviours (smoking, violence, sex, drugs) • Heterosexual peer interests → relationship • Develop vocational plans → Potential financial independence

Guidelines

Recent Royal College of Paediatrician and Child Health (RCPCH) guidelines exist regarding healthcare for adolescents, with specific recommendations for transition, inpatient and outpatient management.¹

Adolescent development and illness

Knowledge of normal adolescent development is essential for managing acute and chronic illness in adolescents. Separate biological, psychological and social strands of adolescent development occur in parallel, with individual variation in the timing in each strand, although biological development tends to occur in early adolescence and biological and psychological developments strongly influence social development. These changes can be split into early, middle and late adolescence but I have amalgamated them for simplification (Table 1).

Biological development

The timing of onset and rate of stages of puberty are variable amongst individuals. Usually girls enter puberty under the control of gonadotrophin releasing hormone, which in turn is controlled by multiple factors, one of which is adequate weight (above 90th percentile of weight for height). They have a growth spurt corresponding with breast

Tanner stage 2 at an average age of 11–12 years; boys' growth spurt occurs at a testicular volume of 8–10 mL, usually at 13–14 years. The timing of puberty influences psychological and social development. There are other physical changes that accompany pubertal growth, for example maturation of organs such as the liver and lung. These changes coincide with the need for extra sleep and it is not uncommon for teenagers to require 12 hours sleep. Acne and blushing may start and be problematic.

Psychological development

Psychological changes in thought patterns and cognitive ability are driven by increasing maturation and myelination of the adolescent brain, as well as pruning of synapses. Children think concretely and can only understand the immediate consequences of their actions and events. From about the age of 12 years the beginnings of abstract or logical thought appear and thought patterns change; adolescents begin to understand cause and effect as well as manipulation of ideas and possibilities for the future. They feel that they are immune to complications (a term that can be described as feeling 'bullet proof') and cannot see the consequences of their actions towards themselves. Eventually they develop complex abstract thinking and are able to manipulate things to help achieve or prevent certain results, such as pregnancy. Adults are not

always better planned but are able to undertake actions that will limit the results of any risk-taking.

Social development

The end-point of social development in adolescence is living as an independent adult. In Western culture, adolescence is becoming increasingly prolonged and often continues into early adulthood. The normal tasks include forming couple relationships, vocational planning resulting in the ability to earn a living and support oneself, and living away from the parental home.

The reciprocal effects of CF on adolescent development

Adolescence can be difficult for healthy individuals, with a spectrum of normal variation in addition to the dynamic process (Table 2). However, chronic illness can disrupt development, and developmental changes may markedly affect the control and management of certain diseases. Illness separates young people from their peers and cloisters them with their parents at the very time they need to be gaining autonomy. Conversely, increased independence when abstract thinking is poorly developed and experimentation is normal may adversely affect illness management through poor adherence. Specific psychosocial areas such as achieving independence and family relationships are vulnerable to dysfunction in adolescents with chronic health conditions. Family connectedness is of fundamental importance to emotional wellbeing.² New diagnoses in middle adolescence, for example diabetes mellitus in addition to CF, are particularly hard to deal with, and additional diagnoses in both early and middle adolescence may provoke concerns about body image and are linked to increased risk of developing an eating disorder.³ Newacheck and Stoodard showed that where more than one chronic disease is present, there is increased morbidity across various measures.⁴

Biological

CF usually results in delayed puberty secondary to a delay in gonadotrophin releasing hormone from the hypothalamus. The delay may be up to two years, with consequent short stature in puberty.⁵ Many CF patients are also underweight due to the increasing catabolic state, poor absorption and production of calorie-dense sputum, as well as the more general effects of chronic disease on eating behaviour.

The vas deferens is absent in 98% of men with CF, rendering them infertile. There is often simultaneous dysfunction of the seminal vesicles, resulting in low ejaculate volume.⁶ CF men thus have normal sexual functioning and potency but small ejaculate volumes. Confusion and poor knowledge surround a range of reproductive and sexual health issues in men with CF. Sawyer⁷ studied the experiences of 50 adolescents and adults: 90% of adults, 60% of adolescents and 50% of parents knew of male infertility in CF. The mean age they first recalled hearing about infertility was 16 years. 48% of adults and 83% of adolescents heard about the issue from healthcare providers. 90% reported no major distress on first hearing about infertility during adolescence. 30% reported an increasing significance of infertility with maturity. 40% knew about small volume ejaculate, but none had been told this by a healthcare provider. 20% confused infertility with impotence. There was agreement by respondents that 14 years is the correct age to initiate discussions, and that semen analysis should be offered to CF men.

Sexual health

60% of CF adolescents report that they want to know the implications for sexual health⁸ before 16 years of age.⁹ The age of onset of sexual activity is reportedly no different than that of their healthy peers, hence they have the same high risk of unplanned pregnancy or sexually transmitted infection as any other adolescent.

Contraception

CF adolescents should be jointly managed by a family planning clinic. The issues to consider include pulmonary hypertension, which is an absolute contraindication to oral contraception, as is symptomatic gall bladder disease. They need specific advice about extra protection when on antibiotics. As with all teenagers, additional use of condoms is advised.

Pregnancy

Due to the risk of passing on the affected gene, it is advised that partners are genetically screened for the cystic fibrosis transmembrane conductance regulator mutation because of the increased risk of CF in offspring.

Men need to know early so they can try assisted contraception if they wish, with surgical retrieval

Table 2
Reciprocal effects of adolescent development and cystic fibrosis

	<i>Effect of CF on development</i>	<i>Effect of development on chronic illness</i>
Biological	<ul style="list-style-type: none"> • Delayed puberty • Short stature • Reduced bone mass accretion • Malnutrition secondary to inadequate intake due to increased caloric requirement of disease or anorexia 	<ul style="list-style-type: none"> • Increased caloric requirement may worsen disease control • Increased insulin resistance at puberty may cause diabetes/worsen diabetic control
Psychological	<ul style="list-style-type: none"> • Infantilization • Adopt sick role • Egocentricity persists later • Impaired development of sense of sexual/attractive self (men) in view of delayed puberty and low weight • Relationship with food affected • Issues around ideal body weight – increased risk of eating disorders • May be prone to depression/anger 	<ul style="list-style-type: none"> • Increasing independence can lead to poor adherence • Poor adherence and disease control due to poor abstract thinking and planning • Self concept of bullet-proof • Reject medics like parents • Risk-taking behaviours
Social	<ul style="list-style-type: none"> • Reduced independence when should be separating from parents • Social isolation (segregation from peers, missing school, illness-related fatigue, medication time, rejected from peers or have fantasies of rejection, admissions, treatment) • Segregation from other CF friends • May lead to vocational failure • identity concerns as future uncertain • Failure of peer and couple relationships 	<ul style="list-style-type: none"> • Associated health risk behaviours • Chaotic eating habits → malnutrition • smoking, alcohol, drug use, sexual risk-taking common and may be exaggerated in view of limited life span

of their own sperm and intracytoplasmic sperm injection (ICSE).¹⁰ Some men reported they would have made this a priority financially if they had known in good time. Women with well-controlled disease will have normal ovulation; if a woman decides to become pregnant she will require careful management, as it places her at greater risk of disease deterioration.

Psychological

There are several different consequences to the correlation of CF and dynamic adolescent change as regards psychological development. Due to usual early diagnosis in CF, the young person has been dependent on their parents for the large treatment burden. This may keep the young person in this child-like, compliant, dependent role for longer than would be expected and disrupt their flow towards autonomy and managing their own illness. This state of co-dependence can become disrupted as the adolescent finally tries to separate from their parents and can cause difficulties in negotiation of the new parent/child relationship. Conversely, the CF patient may become rebellious and reject both medics and parents.

Gender differences

Some studies have reported that girls with CF experience higher morbidity and mortality compared to boys. Girls reported more illness-related strains and worries, including emotional strains, greater treatment discouragement, lower self-esteem, and lower adherence to some aspects of the CF regimen (e.g. coughing, eating high fat foods, taking meds). Living with CF has a greater emotional impact on girls.¹¹

The 'thinness' common in CF can be a positive attribute for young women in their development of a sense of attractiveness and sexual self in western societies. This can also be a problem for men as they are overtaken in height and development by their peers as well as women. This sometimes leads them to overcompensate for their lack of stature in other ways, such as becoming disruptive at school or increasing their risk-taking behaviour in order to fit in with their peer group. Central lines or gastrostomies have a deleterious effect on body image.

Weight and CF

Good nutritional status is linked with improved clinical status and survival. In some conditions

Table 3
Perception of weight in different sex and weights in CF¹³

	<i>Very underweight (see selves as normal/overweight)</i>	<i>Underweight (see selves as normal/overweight)</i>	<i>Normal weight (see selves as underweight)</i>
Girls	29%	41%	19%
Boys	11%	15%	42%

where adequate or correct type of food intake is part of the treatment or eating is linked to the treatment (e.g. creon), eating insufficient calories or the wrong food groups can be another way of non-adhering, or there may be associated secondary gains, such as weight loss to 'improve' the appearance. It is recognized that female adolescents with CF do worse than their male counterparts and find the emotional burden harder. It has also been noted that they have a better body image, as CF tends to keep them slim. They can also adjust their body weight by taking less creon, much as teenagers with diabetes mellitus are reported to omit insulin to reduce their body weight. It is common to have disordered eating in CF, even if the eating habits do not fulfil the full diagnostic criteria for an 'eating disorder'.¹² Bryon and Shearer explored the two worlds that an adolescent with CF inhabits in relation to weight: the non-CF world where weight loss is deemed good and the CF world where weight gain is prized.¹² Walters¹³ looked at young people with CF and assessed weight and attitude to weight in men and women; finding that young women tend to overestimate their weight while young men tend to underestimate it (Table 3).

Perception of underweight was the strongest predictor of taking oral/enteral supplements. 70% of those that saw themselves as underweight were taking supplements versus 30% who did not.

Awareness of limited life span

Adolescence is also the time during which CF patients realize that they have a very limited life span ahead of them and that they will die in their foreseeable future. Friends' or siblings' deaths from CF may bring this realization on suddenly or it may be more gradual. They may adjust by denying the illness and focusing on other things.

Social

School time may be reduced due to disease management, lack of energy and a sense of 'not fitting

in'. Socializing may also be affected by the need to avoid smoke-filled rooms, which exacerbate CF symptoms (e.g. coughing).

The outcome may be reduced social inclusion and hence effects on peer relationships and later sexual relationships, as well as potential vocational failure. In an effort to be like their healthy peers, some CF patients who are able to suppress coughing do so in order not to cough in front of people. Over time, however, this may lead to the patient developing foul sputum, which will affect their breath and kissing.

Experimentation

Comparing CF and sickle cell disease (SCD), chronically ill teenagers report significantly less lifetime and current use of tobacco, marijuana and alcohol, less sexual intercourse, less weapon carrying and more seatbelt wearing than their healthy peers. 21% of CF and 30% of SCD adolescents had smoked and those with CF were less likely to have had early sexual intercourse than those with SCD (28% versus 50%).² In a study by Britto in 1998,¹⁴ significantly fewer teens with CF reported ever smoking or regular tobacco use compared with their healthy peers. Specifically, 21.1% of adolescents with CF reported ever smoking compared with 53.3% of ever-smokers among adolescents without CF matched on age, race, and gender. Almost 3% reported smoking cigarettes for more than two days in the last month, in comparison to 29.6% of regular smokers among adolescents in the healthy comparison group. The median age of onset of smoking (11–12 years) was comparable among adolescents with and without CF. Severity of limitations to health status did not affect how often they engaged in these behaviours.

Coping mechanisms

To cope with chronic illness, the adolescent may adopt one or more of the following coping mechanisms which may or not be in their best interests:

Table 4
Risk and protective factors

<i>Risk factors</i>	<i>Protective factors</i>
<ul style="list-style-type: none"> ● Male sex ● Psychiatric illness or criminality in a parent ● Severe family discord ● Low socioeconomic status 	<ul style="list-style-type: none"> ● Positive temperament ● Above average intelligence ● Social competence ● Supportive relationship with at least one parent ● Family closeness ● Adequate rule setting

- *Insightful acceptance*: this is a healthy and productive response
- *Denial*: this is a common and protective mechanism and has some use but can lead to missed medication and appointments
- *Regression*: taking on increased child-like behaviour and depending on adults
- *Projection*: the adolescent projects his own feelings of anger, frustration, rage or guilt onto healthcare providers, who have to recognize the process and tolerate it or else they may find themselves reflecting these feelings back to the adolescent in an unhealthy spiral
- *Displacement*: this is an extension of projection where the feelings are projected onto an object, and may result in behaviours such as throwing items around
- *Acting out*: this is similar to displacement, and the adolescent may exhibit out-of-control behaviour which can clearly be disruptive in the ward setting
- *Compensation*: this is a highly useful coping strategy whereby the adolescent who previously gained satisfaction and self-esteem from an activity or hobby that is no longer possible with the illness may take up another hobby to replace one they can no longer do in order to gain self-esteem
- *Intellectualization*: some patients become very involved with illness control and the details involved with the technical aspects of care; aspects of this protective mechanism can be useful but the feelings associated also need the time and space to be dealt with.

For those adolescents who cannot continue to cope with the stress and requirement of disease management, coping mechanisms may break down, manifested by poor adherence, risk-taking, withdrawal from developmental tasks and depression. The burden of risk factors and protective factors was looked at by Rutter¹⁵ and later Patter-

son and Blum,¹⁶ who found protective and risk factors important in coping with the burden of a chronic illness (Table 4).

Management of chronic illness in the adolescent

Optimal medical care must be given, with the added exploration of family, development and mental health.¹⁷

The general principles of management are:

- (1) Educate the adolescent and family in the disease and its treatment in developmentally appropriate language
- (2) Respond to emotion – listen carefully and give time to hear their feelings
- (3) Involve the family, because their support is crucial – they must be told to avoid the *over* (over-protection, over-anxiety and over-attention)
- (4) Involve the patient – the more involved the adolescent is in the decisions and treatment, the better
- (5) A multidisciplinary team is essential – nurse specialist, psychologist, dietician, physiotherapist, occupational therapist and social workers must respect each other and discuss the patient's care in interdisciplinary meetings
- (6) Continuity of care – it is helpful to the patient to have an advocate they can trust over time
- (7) Limit-setting if behaviour or adherence is a problem – this avoids splitting
- (8) Support groups – adolescents should be told of support groups even though some will actively avoid these in a desire to feel 'normal' and not identify with the CF population.

You will recall the effects of CF on growth, puberty and development as well as the effects on weight gain. The management of these must involve the multidisciplinary team. Overall

improvement in disease control has the best chance of improving growth and development. Once the disease and malnutrition have been addressed, growth should follow.

Communicating with adolescents

Adolescents are neither children nor adults: they have their own unique needs. Each young person possesses their own beliefs about their health, illness and general life issues which need to be discovered, respected and considered. The main skills needed are the ability to engage and form a relationship with the adolescent in order to understand his situation and manage the illness together in partnership. The adolescent consultation has some major differences from that with a younger child, predominant amongst which is that adolescents have a much larger role in the consultation, a role that will change with continued psychosocial development. There is also the new role of parents to be considered.

What do they want in a health professional?

An effort to make healthcare for adolescents more patient-centred must be grounded in adolescent opinions. Most important in Britto's study¹⁸ were physician trust and respect, patient power and control, and caring and closeness in the doctor-patient relationship. They preferred communication to be directed at them rather than at their parents. They rated honesty and attention to pain, and valued both scientific as well as interpersonal aspects of care.¹⁸

Adolescents want a competent adult who they respect for advice. Klostermann¹⁹ compared those with and without chronic illness with respect to how they perceived doctor-patient trust. They rated competency, fidelity, confidentiality, honesty and a global perspective. Younger adolescents are more concerned with confidentiality and adolescents with chronic disease are more interested in involving their parents in their care. Specific behaviours that improve trust are asking for adolescents' opinions, keeping information private, not withholding information and engaging in small talk to show concern.

Adolescent consultation

Practicalities

Although the central relationship is with the adolescent, the family is crucial to successful support and management of a chronic illness. There are

several models of consultation, including seeing the family alone, then the adolescent alone, then all together at the end, and seeing the family together then the adolescent alone, then together again at the end.¹⁷ It is essential to prove confidentiality not to see the parents alone after you have seen the adolescent alone.

Longer times are needed for adolescent consultations and afternoon or evening clinics allow them to attend as much school as possible.

Healthcare profession gender

50% of girls prefer a female provider and 23% boys prefer a male provider. During examination most younger girls wanted a parent present, younger boys had no preference. Girls with a female physician are more likely to have private time. Only 50–60% had spoken privately with their doctor.²⁰

Confidentiality

Confidentiality and consent issues are central to the management and examination of young people, who are potentially legally underage. Adolescents are very clear that the major things they want from clinicians are confidentiality, respect and clinical excellence. Full confidentiality (including keeping confidentiality from parents) should be assured to young people unless they are found to be at risk from suicide or sexual abuse, or reveal plans to harm others.

Consent

In relation to issues of consent to treatment, adolescents can fall into a no-man's land between parental rights over minors and adult rights. In most countries, including the UK, adolescents are now deemed to have adult rights to consent to treatment themselves if they are legally competent, regardless of their parents' wishes. The legal criteria for competence differs between countries, but usually requires the ability to give informed consent and understand the benefits and risks of treatment or non-treatment. In the UK, competence is presumed over the age of 18 years, and adolescents between 16 and 18 years can consent to treatment but cannot refuse life-saving treatment. Under 16 years of age, adolescents are legally presumed incompetent unless they show otherwise. Nonetheless, many young people under this age are competent, so in a practical sense it is appropriate to treat adolescents from 12–14 years upwards as if they have full adult medical rights and responsibilities.

Table 5
HEADSS questions for history taking

<i>Heading</i>		<i>Questions</i>
H	Home life	Relationships, social support, household chores
E	Education	School, exams, work experience, career, university, financial issues
A	Activities	Peers, who can they rely on? Exercise and sport
D	Driving	Aged 16 if disabled
	Drugs	Cigarettes, alcohol, how much? How often?
	Diet	Calcium, vit D, Weight, Caffeine (diet drinks), binges/vomits
S	Sex	Concerns, periods, contraception (and in relation to medication)
	Sleep	How much? Hard to get to sleep? Wake often? Early waking?
	Suicide	Depression, disabled adolescent men high risk, ask about mood

Communication principles

In order to form an alliance with an adolescent they need to feel you are interested and care. Open-ended questions are the most useful in drawing out a reserved adolescent. Instead of asking, for example, 'Do you like school?', it may be more revealing to ask what the best parts are about school or what they dislike about it. When trying to uncover issues such as bullying, use questions such as 'We know that at a lot of schools, some people get picked on – does that ever happen to people you know at your school?' before narrowing it down to the person themselves. Other useful questions include, 'How is family life?', 'How do you get on with your friends?', 'Where do you go when you go out with your friends?', and 'What sort of things do you get into arguments with your

parents over?' When considering adherence it is essential to normalize it, as we know over half of patients will be non-adherent to each medication. Some helpful questions may be 'We know how difficult it is to do all the physio and take all the medicines you have been prescribed to look after your CF. What would be useful to know is how much have you actually managed to do over the past week.', 'Which of your treatments are the easiest to take and which are the hardest', and 'What sorts of occasions/reasons make it harder to do your physio?' The main points to help with adolescent consultation are shown in Box 1.

Additional questions

It is important to get used to asking questions specific to an adolescent's life in an increasingly probing order so that any issues can be easily mentioned. HEADS (Home life, Education, Activities, Driving, Drugs, Diet, Sex, Sleep, Suicide) is a useful acronym to follow (Table 5); however, questions must be appropriate to the young person's stage of development. It is important to use this on a regular basis since it has been shown that young people have health concerns in the areas of sex, smoking and mood that they will only address if specifically asked.

Examining young people

Adolescents require more attention to privacy and confidentiality during examination than children. Ensuring personal privacy is essential, and it is appropriate to ask young people whether or not they wish their parents to be present during physical examination. Be sensitive to the fact that they may not wish their parents to be present but may have difficulty saying this in front of parents. This can be dealt with by suggesting to the parents that

Box 1 **Essential issues in adolescent consultation**

See adolescent alone for part of consultation
Confidentiality and consent
Take your time
Consider late afternoon times
Engage the adolescent as the central person in the consultation
Use non-judgmental questioning about risk-taking behaviour and be curious
Use non-judgmental questioning about adherence
Be frank, and avoid authoritarian approach
Respect confidentiality
Remember proxy presentations are common as an excuse to see the doctor
HEADSS questions
Assess what they have remembered from previous earlier discussions
Explain why you need to examine them. Do they want a parent present or same-sex doctor?
Explain your findings in developmentally appropriate language

the young person may now be old enough to want privacy and then asking the young person what is preferred.

The issue of the gender of the doctor and chaperones is important. Many but not all adolescents prefer to be examined by a same-sex doctor, and providing a gender balance and choice of examiner is useful if this is possible. Having a chaperone for examination of adolescents of the opposite sex is mandatory to protect both the patient and the doctor.

Pubertal assessment

Assessment of pubertal stage is important for the management of chronic illnesses in adolescents, as it is related to growth and weight. Pubertal stage should be assessed at least annually in young people with chronic illnesses during early adolescence. For those who refuse direct genital examination, pubertal self-assessment using standard Tanner pictures is an option.

Treatment burden in CF

It is well recognized that the treatment regimen in CF is both time-consuming and unpleasant, as well as relentless over the patient's lifetime. It also becomes more onerous with time and decreasing lung function. In childhood those who adhere well may be able to delay the inevitable decline. If they are already noncompliant as children they may have deteriorating lung function in adolescence and be symptomatic, requiring a regimen to keep sputum at bay and coughing suppressed during the day. When illness is well under control then treatments are not felt to give an immediate response. As the illness and respiratory function deteriorate, however, treatment has more immediate benefits such as preventing coughing later.

Patients are expected to do physiotherapy (physio) twice a day with bronchodilation and DNase before physio as well as nebulized antibiotics and sometimes steroids. They may also be on oral or intravenous antibiotics, oral creon and energy drinks and vitamin supplements. They are also required to exercise frequently as part of their treatment.

If they develop diabetes then there is a significant further burden which can hit hard, especially during adolescence.

Ziaian²¹ compared treatment in CF compared with diabetes and asthma and found that the average time spent was 74.6 ± 57 minutes in CF versus 56.9 ± 27.8 minutes in diabetes.

Adherence

Adherence is defined as the extent to which a person's behaviour (in terms of medication, following diets or executing lifestyle changes) coincides with medical or health advice.²²

Adherence is a particular issue in young people as they are beginning to take over management of their illness. In addition, certain developmental issues (e.g. abstract thinking capacity) influence the ability to adhere strongly, and young people often prioritize management of their illness behind social and recreational activities in a bid to feel like their healthy peers.

Adherence rates

It is well recognized that adherence in chronic diseases runs at about 50%.²³ Poor adherence to medical regimens in chronic disease is common in children and adults as well as adolescents.

Modi *et al.*²² showed that less than 50% of children under 13 years old with CF adhered to treatment, with differing rate depending on treatment component. Zindani *et al.*²⁴ showed slightly higher rates of 57% for multivitamins and 78% for a mucolytic nebulized medication (dornase alpha) using the medication event monitoring system Smart-Caps. Adherence rates for chest physio are about 40–47%,²³ while adherence to dietary recommendations are lower, ranging from 16–20%.²⁵

Only 29% of adults undergo daily chest physio and their reasons include difficulty fitting it into their lifestyle, a perception that it does not help, the physical consequences of chest physio, doing exercises instead, and doing it as and when necessary.²⁶

There is a clear association between poor adherence and unfavourable transplant outcome. Bullington's paper identified three main reasons for medication non-adherence following transplant, namely medication issues, troubled adolescence and deliberate non-adherence.²⁷

Adherence varies with treatment component

It is essential to remember that adolescents may adhere differently to different aspects of management. Modi *et al.*²² looked at different assessments of adherence, including child and parent telephone calls and electronic data as well as diaries. Diaries correlated more closely to electronic data and parent/child reporting was often inflated.

Reasons for poor adherence

There are many reasons for non-adherence, as described below. It is essential to remember that it is not the patient that is the problem but the non-adherence, and that this may be multi-factorial.

In 2003, the World Health Organization²⁸ described the five pillars of non-adherence as follows:

- Patient related
- Social and economic
- Healthcare system
- Condition related
- Therapy related.

Patient related

Patient related non-adherence may be part of the adolescent rebellion or secondary to denial of the illness.

Adolescents with poorly developed abstract thinking tend to see themselves in the present and are less able to understand the future consequences of their actions. As future planning capacities develop, young people often tend to see themselves as 'bullet-proof' (i.e. that problems will only affect others).

Adolescents prioritize their social life ahead of illness and hence do what they can to feel like their friends without CF. They may try to get away with the minimum of treatment to feel like their healthy peers, and not stand out. They may have incorrect beliefs.

Social and economic

There are also family reasons that impact on adherence. Earlier treatment will have been led by the parents, and they may have got into relationship difficulties with the adolescent around treatment or in general. Adolescence is the time of starting to separate from being a dependent child and they may reject parental values and good behaviour and rebel before they are ready to make those decisions.

Other reasons are lack of parental support, limited English language, lack of family or social support networks, the burden of the regimen, difficulty accessing treatment (e.g. transport, distance), cultural beliefs about health and illness, or a chaotic home life.

Healthcare system

In some cases knowledge plays a part, as some young people may not have learned about their

illness and how medications work as they make the transition to adolescence. There is some evidence that families that are less knowledgeable can be less adherent to regimens.²⁹ Once basic knowledge is there, improving education does not improve adherence. Other factors are the doctor-patient relationship, healthcare provider communication skills, health belief disparity between the patient and provider, lack of education/explanation of health knowledge, and lack of continuity of care.

Condition related

Adherence is affected by illness chronicity, severity, lack of symptoms, and future consequences of disease.

Therapy related

Adherence is also related to the complexity of regime, whether treatment requires mastery (e.g. inhalers, injection), duration of therapy, frequent changes, lack of immediate/observable benefit, actual or perceived unpleasant side effects/stigma, and significant interference with daily life. Patients may have unpredictable beliefs about their treatments. During in-depth interviews with 32 7- to 17-year-olds, Williams³⁰ explored the difficulties with chest physio and found that it was frequently described as restrictive, threatening to identity and boring, giving rise to feelings of unfairness, inequality, difference and social stigma. Motivation to adhere was influenced by perceptions of effectiveness that depended on external signs evident during or after the physio. Visualization of mucus accumulation enhanced motivation. Distraction techniques reduced the perception of duration. This is a common theme in that treatment that is noted to be immediately helpful is easier to adhere to as there is a clear benefit.

Sometimes there are practical barriers to adherence, such as not liking the taste of a medicine.

Barriers to adherence

Modi *et al.*³¹ looked at specific barriers to adherence and found that forgetting, oppositional behaviours and time management problems were frequently mentioned by young people. As expected, more barriers were associated with poorer adherence. The more adherence points the worse the adherence.

Improving adherence

Engage

The main objective is to engage and form a relationship built on honesty and trust, where doctor and patient can work together. It is recognized that most adolescents actually do adhere to a regimen of their own choosing according to their own beliefs and priorities. Our task is to find out what that is.

Determine the patient's perspective

It is crucial to determine how the patient perceives his disease: something to be denied and hidden, or to be accepted and treated.

Determine the level of adherence

Modi²² suggests that normalizing non-adherence should be done at all clinic visits. This will allow non-adherence to be worked on, with further referral for behavioural or psychological techniques if necessary. An example of how to start the conversation could be 'Most young people find it difficult to do all of their different treatments every day; we'd like to know what you are currently managing to do on a daily basis'. Asking questions that apply to a specific timescale is also recommended, for example, 'Over the past week, how many times have you managed to xxx?'. In order to improve adherence the initial task is to quantify the level of adherence for each treatment.²²

Determine the barriers to adherence

These can be anything from simply not liking the taste of a particular medicine, to family reasons, to not prioritizing illness management to rebellion. Only then can the reasons be worked on. It is useful to consider and ask what the barriers are. Peer relationships and self-image are very important: for example, taking the latest trend in bags to school may be more important for an adolescent with CF than whether or not the bag is big enough to carry their medication, and going to see the school nurse for tablets would mean not sitting with friends in the canteen. Side-effects are also important, particularly those that may affect well-being or appearance, such as weight gain with steroids.

Once you know the barriers you can work together on each one. You are there for advice and can negotiate. It is imperative to involve family as well, since they are the ones living with the

condition and the adolescent and can gently nag (Table 6).

The multidisciplinary team

The members of the multidisciplinary team are essential in good care of young people. The clinical nurse specialist, nurses, physiotherapist, dietician, psychologist, activity coordinators, teachers, psychotherapist, child and adolescent psychiatrists, consultant and junior doctors may all have a place in the treatment. Patients should be discussed between the team with all parties' contributions equally valued. If patients are noticed to be trying to split the team, this is important information to note and the team must try to stand united. Psychological support is valuable in these situations for both staff and patient.

Inpatients

The Bridging the Gaps publication by RCPCH addresses secondary care and has some points to be adhered to:¹

- Every hospital should have a policy regarding the care of young people in hospital
- At the minimum, they should be looked after in a separate facility on a children's or adult ward, preferably of mixed gender
- The ward should be managed by a nurse and doctor with responsibility for young people to ensure standards of confidentiality, consent and privacy
- Nursing staff must include a lead appropriately qualified in adolescent mental health
- There should be appropriate educational facilities and transition to employment schemes
- There should be emergency access to child and adolescent mental health services (CAMHS) as well as CAMHS liaison.

Transition

The Department of Health published a document called *Transition: Getting it Right for Young People*.³² Transition is an essential component of high-quality healthcare in adolescence and every paediatric general and speciality clinic should have a specific transition policy. Paediatric and adult models of outpatient care differ and the young person will probably find that once transitioned to adult services they will be seen less frequently. Transition is not age-related. Young

Table 6
How to improve adherence

<i>What</i>	<i>Why</i>
Engage	Take time to see situation through their eyes
Motivation	Search for factors that they actually care about (e.g. puberty or growing which will start once disease control is improved)
Assess the size of the problem and decriminalize	Ask: 'most people have trouble taking their medication. How often do you remember yours?'
Involve	Plan the regimen with the adolescent
Take time to explore practicalities	If you put yourself in the adolescent's shoes with a mind for detail, you may find the problem – sometimes it is as simple as not having a bag big enough to carry the medicines around in
Think practically	Find an activity they do daily and attach the medication to it (e.g. brushing teeth); find the least chaotic time of day, often the morning
Visualize	It is easier to remember to do something if you can visualize yourself doing it (e.g. taking pills 'with breakfast and dinner' instead of 'twice a day')
Find out what they would agree to do	You then have a place to start working together from
Barriers	Ask about each drug in turn and explore barriers to taking, and what makes it hard
Contract	Make a contract that they agree to
Written instructions	Something to remind them – most of what is said has been shown to be forgotten once they leave the room
Take time to explain	It is a good idea to check out their level of knowledge on each occasion
Explore beliefs	You may be surprised to learn about their beliefs (e.g. that one drug works perfectly but they don't think the other does [or don't like the assumed side effects] so they only take the one)
Solution-focused approach	Find out what has been going well and why (e.g. ask 'How have you managed to stay well for three weeks this month?'), and use this information
Check motivation	See where they are in the motivational cycle and if pre-contemplative you may want to refer to psychology to work on motivation

people should be transferred to adult services when they have the necessary skills to function in an adult service and have finished growth and puberty. Some points to note are:

- Preparation for transition should start early, as it takes time to get to know and trust the new team
- A transition clinic can be useful if there are large numbers of transition patients; it is usual that on each occasion the young person sees a familiar doctor or nurse as well as meeting the members of the new team
- An identified person within the paediatric and adult teams must be responsible for transition arrangements; the most suitable persons are nurse specialists
- Management links must be developed between the two services, and local commissioners must be consulted when patients are transferred from one tertiary centre to another.

Summary

The treatment of adolescents with CF is important, as adolescence is often when the CF patient's health deteriorates. They are managed by paediatricians then transitioned to adult teams. For the person with CF, adolescence usually constitutes a major chunk of their life span. The treatment is set on a background of dynamic developmental change. This can cause specific difficulties in management, especially related to eating and adherence level. Developing good relationships with young people, seeing things from their side and seeing them alone every time as well as with their families will help. They may have health-related issues to discuss with you and should be asked HEADS questions regularly. The multidisciplinary team is essential when working with young people. With the right approach, working with young people can be very enjoyable and productive to both parties.

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