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Diagnosing autism

Kirsty Yates

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Abstract

Interest and awareness of autism within public and medical domains has increased over recent years. Parental awareness has resulted in children being presented for earlier advice, assessment and diagnosis. Routes of entry for referrals can vary due to the diversity of the condition and local service provision. It is therefore necessary for paediatricians, general practitioners and allied professionals with a contributory role within developmental child health to have a sound knowledge of the presentation and assessment of autism spectrum disorders (ASD). Early identification of deviation away from the typical developmental trajectory in this group of patients is advantageous in order to maximize their potential, provide targeted intervention and minimize co-morbidities. This review addresses the diagnosis of ASD and provides an assessment framework for professionals who encounter a child with suspected autism.

Keywords asperger syndrome; assessment; autism; autism spectrum disorder; diagnosis; pervasive developmental disorder

What is autism?

Autism is a neurodevelopmental disorder, classified under the umbrella of conditions known as pervasive developmental disorders.^{1,2} The term autism refers to the prototypical condition described in 1943 by Leo Kanner and is also known as core autism or early infantile autism, due to its presence in early childhood. It is characterized by qualitative behavioural abnormalities in the domains of communication, reciprocal social interaction and interests, and activities that are repetitive, restricted and stereotyped. The abnormalities are pervasive and therefore present across different settings. It is recognized that there is a spectrum of the condition and a broader autism phenotype with less severe and more subtle behavioural features. Although not recognized by the DSM-IV TR and ICD-10 classifications as a specific disorder, the term autism spectrum disorder (ASD) is used both in lay and medical terms to encompass the broader condition and includes autism, Asperger syndrome, pervasive developmental

disorders not otherwise specified (PDD-NOS) and childhood disintegrative disorder.

Epidemiology

ASD is not rare. Recent replicated prevalence rates report estimates of 30 per 10 000 for core autism and up to 1 in 100 for ASD.³ The condition is three to four times more common in boys, with a male preponderance rising in the high functioning group.⁴ Over the years epidemiological studies suggest prevalence has been increasing. This is likely at least in part to the broadening of the diagnostic criteria and inclusion of cases previously classified differently, along with improved case recognition.³⁻⁶

What causes autism and autism spectrum disorders?

ASD is now accepted to be a neurodevelopmental condition with a biological basis. Despite increasing research, no clear aetiology has been identified. The heterogeneity of affected individuals and genetic complexity has undoubtedly contributed to the daunting task of identifying the cause(s) of ASD. The condition is likely to be multifactorial in origin with genetic heritability and environmental factors possibly influencing phenotypic expression.

Taking into account twin studies and the population base rate for autism, genetic heritability of core autism is approximately 90% with a recurrence risk in siblings of 6%.⁷ It is possible that several genes of small effect may act through an epigenetic mechanism,⁴ and even though multiple susceptibility genes have been identified, few of these findings have been replicated. Chromosomes 2, 7 and 16 have been consistently cited and cytogenetic anomalies on chromosomes 15 and 22 have also been reported.^{6,8}

In a minority of cases (less than 10%),^{4,5,7} ASD is associated with a known medical condition, but mechanisms relating to causality are not well understood. Consistently recognized genetic conditions include tuberous sclerosis (TS) and fragile X. Studies have shown that between 1% and 3% of children with autism have TS and similar percentages have fragile X.^{4,7}

There is an increasing body of research studying neurobiological differences in ASD compared with controls, looking at variation in neurotransmitters, and volumetric and functional differences of various regions within the brain, but further studies are needed to ascertain relevance for clinical practice.⁸

Clinical research has also demonstrated differences in trajectories of head growth in children with ASD.⁶ Macrocephaly is a recognized feature of ASD in 20–30% of cases.⁴ Studies have shown that as a group, head circumference accelerates during the first 2 years of life, with deceleration possibly occurring in later childhood since average head circumference has been reported in adolescence and adulthood.⁴ Although there have been conflicting views around the relevance and cause of these changes, they are reported to happen prior to the onset of clinical symptoms and therefore may be a useful clinical indicator.^{6,8}

Various environmental factors have been reported in the literature. Prenatal influences, including maternal alcohol and valproate use and hypothyroidism, have been linked to autism as have congenital infections, but congenital rubella is the only established link.⁷ There has been much controversy following links between the MMR vaccine and a so-called regressive type

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of autism. Evidence consistently states there is no causal link between the two, and that the hypothesis of the vaccine causing a gut disorder leading to autism is unfounded.^{7,9,10} Epidemiological studies have also shown that links between thimerosal, a mercury containing vaccine preservative no longer in use, and autism are again unfounded.^{4,7}

Making a diagnosis

ASD is a heterogeneous condition with no single pathognomonic feature. Diagnosis can be challenging as affected individuals can not only display variation in the degree of behavioural severity, language and intellectual abilities across the three developmental domains, but their behavioural profiles can change with age. For a diagnosis of core autism, ICD classification² states that abnormal or impaired development should be present by the age of 3 years. Many parents express concerns as early as 15–18 months of age.⁵ Despite this, average age at diagnosis is 4–5 years,^{6,10} possibly due to a combination of factors that include lack of recognition of subtle difficulties at a young age, variability of assessment pathways and broadening of the spectrum to include higher functioning individuals who tend to present later. For example, a pre-school child may activate a referral due to concerns with their speech, whereas a child who is high functioning and has adequate language skills may present later with difficulties in peer interaction becoming more apparent as academic and social demands increase.

Behavioural features of autism spectrum disorders^{1,2,4,5,9–12}

Reciprocal social interaction

Difficulties and delay in social interaction are often the earliest features in ASDs, but they can be subtle and easily missed. Absence of joint attention (i.e. failure to show interest, share a focus of attention and follow gaze) is highly suggestive of core autism. Carers may describe that the child fails to respond to their name when called repeatedly, raising the possibility of a hearing impairment. Inadequate facial expressions, including lack of social smiling and limited use of gestures, e.g. shaking head, nodding, waving, clapping, are also features. Individuals with ASD lack awareness of others feelings and the impact of their behaviour on others. Sometimes this manifests as inappropriate behaviour in a specific social context or inappropriate response to others' emotions. There can be misinterpretation of tone of voice and facial expressions of others, leading to difficulties with peers, often combined with the failure to develop mutual sharing of interests, activities and emotions. Younger children may not seek to share enjoyment, e.g. showing a toy to a parent or pointing out objects of interest to others. Conversely, higher functioning individuals often seek interaction with others and make attempts to socialize but come across as socially odd. Often, social play is limited and in isolation to their peers.

Communication

Concerns may be raised when a child has failed to acquire language as expected. Some children with autism may develop no useful communicative speech or sounds. In contrast to those with specific language disorders, children with ASD often fail to use

gestures or mime to compensate. Instead, parents may describe the child either obtaining a required object themselves or taking another person's hand to the object as if to use them as a 'tool'. Language is often atypical with idiosyncratic use of words or phrases, e.g. nonsense or jargon words, or referral to self as 'you' (pronominal reversal). Other features include delayed echolalia and abnormal delivery of speech (prosody), i.e. unusual pitch, speed, volume or tone. For example, children may speak in a monotonous or sing-song voice or use stereotyped phrases (with constant form or pattern). Delayed echolalia is the term applied to copied or directly imitated speech, e.g. from an adult, television or radio, that is repeated some time after it is originally heard. Whatever the language skills present, two-way reciprocal conversational interchanges tend to be difficult, particularly if the topic of conversation is narrowed and repetitive. An individual with ASD often struggles to engage in social chat and build on conversation outside their area of interest with someone else.

Interests and activities

In addition to limitations in make believe and pretend play, interests and activities in individuals with ASD are often restrictive and repetitive. A child may have a preoccupation with an interest that is abnormal in intensity, content or both, e.g. shiny objects, traffic lights or a non-functional aspect of a toy such as the noise or vibration it generates. For many individuals, play may lack creativity and imagination, but isolated examples of pretend play and imitative behaviour do not exclude a diagnosis of ASD.^{9,10} Change in routines or environment are often resisted, not uncommonly resulting in temper tantrums. Stereotypies or mannerisms are commonly seen but are not always evident before the age of 3 years.^{4,10} Stereotypies in more severely affected individuals can include hand flapping, finger flicking, rocking, head banging and twirling. Sensorimotor interests may also feature in the presentation. Hypo- or hyper-sensitivity to environmental stimuli is often recognized, e.g. loud noises, insensitivity to pain, or fascination with smells, textures or colours of food or fabrics. Some individuals with ASD have superior or splinter skills/giftedness in one or more aspects, e.g. calculations, memory.⁴

Regression

Depending on the sample studied, regression occurs in 10–30% of cases.^{4,5,9,10} Regression most commonly affects language, usually at the less than 10 word stage, therefore occurring between 18 and 24 months of age. Other skills can be affected and parents may concurrently report a change in sleeping or eating habits, loss of eye contact and development of a specific interest.¹⁰ Regression can also occur in children with ASD above 24 months,⁵ but pre-existing development is usually atypical.^{4,12} Any child who presents with features of regression warrants careful medical assessment, including consideration of neurodegenerative conditions, such as Rett syndrome and Landau Kleffner.

Learning disabilities

Historically, autism was only recognized in individuals with severe impairment and learning disabilities.⁷ Estimates suggested learning disabilities (IQ less than 70) affected 70–80% of individuals with ASD,^{4,5} although over recent years and with widening of the spectrum to include higher functioning individuals, figures of 40–50% have been quoted.^{4,8,9} Those with high

functioning ASD may show good verbal skills in contrast to those with autism or PDD-NOS who may perform better in non-verbal measures. However, this 'higher' degree of intelligibility on performance tests cannot be assumed to reflect their social skills which may be significantly impaired.^{6,11}

Epilepsy

The risk of epilepsy in ASD is linked to lower IQ, with peaks of incidence occurring at pre-school age and adolescence.^{8,9} Between 18% and 29% of children with ASD are affected and any seizure type can occur. Epileptiform EEGs are common in autism, both with and without regression,^{10,11} and studies have shown that 10% of children with autism have an epileptiform EEG without any clinical evidence of seizures.⁸ There is no evidence that these discharges have a causal relationship to ASD or that routine EEGs should be performed. Any investigation and treatment should be guided by the clinical presentation of the individual.

Psychiatric, neurodevelopmental and behavioural co-morbidities

Disturbances of behaviour, attention, activity, thought and emotion are common in children with ASD and/or developmental difficulties.¹³ In fact, children with ASD can have any developmental, medical and mental health condition experienced by children without ASD.¹² Disordered sleep and food selectivity are well recognized.^{5,10} Problematic emotional reactions and behaviours can occur and may include self-injurious behaviour, aggressiveness, temper tantrums and emotional lability. Co-morbidities within ASD are well recognized and have been reported to affect up to 72% of cases, with recent studies highlighting the importance of identifying co-morbid mental health problems, particularly in those with higher functioning autism.¹³ A list of psychiatric and neurodevelopment disorders associated with ASD is given in Table 1.

Assessment

The purpose of assessment in ASD is to make a diagnosis where applicable and to guide interventions and treatment based on the needs of the child and family. This includes identification

of co-morbidities and associated developmental problems which can have significant impact on a child with ASD and their family. Diagnosis may be difficult due to communication impairment and possible associated cognitive problems, making it hard to determine whether the features are the result of ASD, due to co-morbid conditions or environmental factors, or a combination of all three. Studies have shown that diagnosis of autism at 2 years of age is possible and stable over time,^{10,14} although it is less reliable for the broader spectrum.

History

The diagnosis of autism is based on history and observation in several settings, looking for core behavioural features. Although screening tools have been used previously, their universal use is not recommended.^{5,11,12,15} Diagnosis should be based on clinical judgement and expertise. Standardized instruments may enhance or facilitate diagnosis in that they can bring a broader understanding to the strengths and difficulties experienced by the patient and family, but they are not essential for every assessment, should not be used in isolation and are less reliable in the younger age group (less than 2 years).¹⁰

Any concerns highlighted by parents/carers or during routine developmental surveillance require further assessment. Key features indicating further evaluation is essential are absence of babble, gesture or pointing by 12 months, no single words by 18 months, no two-word spontaneous phrase (non-echoed) by 24 months and any loss of language or social skills at any age.^{4,5}

Referral entry for a pre-school child will tend to be through child health, e.g. presentation with developmental delay, speech and language difficulties. The entry point is more diverse in school-age children as they can present with a broader range of symptoms, e.g. behavioural difficulties or co-morbidities.

Certainly any deviation away from the typical developmental trajectory warrants a general developmental assessment. It is important to note that a lack of concern from the parents about early development does not imply a normal developmental history.

The differential diagnosis for ASD and associated medical conditions is listed in Table 2, and should help guide further assessment. Even though there are small scale studies in the literature reporting high rates of gastrointestinal symptomatology in children with ASD, there is a lack of published rigorous data to support this finding.¹¹ Nevertheless, symptoms of gastrointestinal dysfunction should be sought in the history in addition to identifying feeding and sleeping problems. Differentiating global developmental delay (GDD) from ASD can be done by taking an ASD-specific history to elicit the core features that are more likely to be out of keeping with the general developmental level of the child in ASD.¹⁰ Determining ASD with learning disabilities from GDD is a more difficult challenge. The 'aide-memoire' in the National Autism Plan for Children (NAPC)¹¹ can be used as a proforma to provide an ASD-specific developmental history. Information should also be gathered from other settings including home, social services (if applicable) and education.

Examination

General examination should be performed as part of the developmental assessment and include a full neurological examination,

Psychiatric, behavioural and neurodevelopmental co-morbidities associated with autistic spectrum disorder

- Attention-deficit hyperactivity disorder (ADHD)
- Tourette syndrome/tic disorder
- Dyspraxia/developmental coordination disorder (DCD)
- Dyslexia
- Obsessive-compulsive disorder (OCD)
- Specific phobias
- Anxiety
- Depression/mood disorder
- Sleeping difficulties
- Feeding difficulties

Table 1

Differential diagnosis of autism spectrum disorder (ASD)

- Global developmental delay
 - Learning difficulties
 - Hearing problems
 - Visual impairment
 - Specific Language disorders
 - Selective mutism
 - Reactive attachment disorder
 - Lack of opportunity for interaction
 - Rett syndrome (if features of regression)
- Some medical conditions associated with ASD:
- Tuberous sclerosis
 - Fragile X
 - Down syndrome
 - Neurofibromatosis
 - Phenylketonuria (untreated)
 - Fetal alcohol syndrome
 - Smith-Lemli-Opitz syndrome
 - CHARGE syndrome
 - Duchenne muscular dystrophy
 - Congenital rubella
 - Iron-deficiency anaemia

Table 2

looking for neurocutaneous stigmata and dysmorphisms, and including Woods light examination. Observation of behaviour in different settings, e.g. home and school/nursery/play group, will allow assessment of the child in environments with varied structures and give an impression of how the child interacts with peers and adults and how they adapt to predictable and less predictable routines.

Investigations

Investigations should aim to exclude or identify medical conditions associated with ASD, but should only be performed where there is specific management, treatment or genetic implications. Paediatricians should be aware of the evidence base as to which investigations are appropriate, and ensure parents receive peer-reviewed and consensus driven information as to why a particular test is or is not indicated for their child.^{4,9}

Investigative yield is generally low, quoted as between 8% and 37% depending on the population studied. Yield of positive investigations is increased if there is lower IQ and dysmorphic features.⁸ Conversely, presence of autism in a cohort of children with GDD and learning disabilities has been shown to decrease the chance of a positive yield.⁴

Karyotype and fragile X testing should be performed in all individuals, with other investigations guided by clinical presentation and family history. Neuroimaging and EEGs should only be performed if there is a clinical indication. Similarly, evaluation of the gastrointestinal tract should be guided by clinical presentation and should follow standard evaluation as for any child.¹² Hearing and visual impairments should be ruled out by performing appropriate assessments.

Multiagency assessment

Referral for multiagency assessment should be made if ASD is suspected after the general developmental assessment. It should identify ASD, learning disabilities and co-morbidities, but also needs to be skills based, aiming to determine the patients' individual profile.¹² Previously obtained information should be collated, in addition to performing cognitive, family and speech and language assessments. This will involve a team of professionals that may include child psychiatry, paediatrics, speech and language therapy, psychology, occupational therapy and physiotherapy. Multiagency work requires careful coordination and this will undoubtedly impact on assessment timescales. The NAPC¹¹ recommends a maximum of 30 weeks to completion of multi-disciplinary assessment from the time concerns are first raised. Standardized history and observation tools, including the Autism Diagnostic Interview – Revised (ADI-R), the Diagnostic Interview for Social and Communication Disorders (DISCO) and the Autism Diagnostic Observation Schedule (ADOS),^{11,12} are likely to be used but require specific training. In some cases referral for regional tertiary assessment is needed when there is diagnostic uncertainty or complexity.¹¹ Regular feedback to the parents is an essential part of the process at all levels of assessment.

Management

Management should be multidisciplinary and take a behavioural and educational approach, as well as providing information and support to the family in the form of a family care plan.¹¹ Identification of strengths and difficulties should generate access to targeted skills-based interventions for the family and child, even if there is diagnostic uncertainty. Evidence is emerging that early social and communication based intervention programmes are effective,^{4,9,10–12,15} including parental group training such as the 'More than Words' and 'EarlyBird' programmes. Educational needs should be met with appropriate support and resources in place, e.g. use of visual augmentation for communication such as the picture exchange system (PECS) and the TEACCH (Treatment and Education of Autistic and related Communication Handicapped Children) approach. Families will require information on local support groups and agencies; therefore, knowledge of local services is required. Referral for genetic advice may also be necessary to discuss implications for future or current siblings.⁴ Identification of an associated medical disorder or co-morbidity should result in management for that condition by the appropriate professionals. Use of medication should not be withheld because of an underlying ASD and may include stimulant medication for ADHD, melatonin for sleep disorders, selective serotonin reuptake inhibitors for obsessive-compulsive disorder or ritualistic behaviours, and risperidone for aggression and irritability, although careful monitoring is warranted.^{11,12,15} Follow-up of developmental progress, changing needs of the patient and additional medical problems are required. Although anecdotal evidence exists, current levels of scientific evidence are insufficient to support the use of biomedical interventions in the management of ASD, e.g. dietary supplementation and gluten- and casein-free diets.^{8,11,12} It therefore remains a controversial area with pressure, particularly from parental groups, for further research.

Prognosis

ASDs are lifelong neurodevelopmental conditions. Behaviours and presentation vary over time with a tendency for progress in all domains, although there is huge individual variation. Determinants of outcome include severity of behaviours, cognitive abilities and useful speech.^{4,12} Many individuals require specific supports; some adults with higher functioning ASD may be able to live independently and obtain employment but at the present time few adults appear to achieve their full potential. This may be a reflection of lack of targeted early intervention in this group, though recent studies have shown those receiving early intervention focusing on skills development have a better outcome.^{4,5,14,15}

Conflict of interest

ALC is one of the authors of the ADI-R.



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FURTHER READING AND USEFUL RESOURCES

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www.cafamily.org.uk Contact a Family.

www.nas.org.uk National Autistic Society.

www.researchautism.net Research Autism.

Practice points

- ASD is a heterogeneous lifelong neurodevelopmental condition with behavioural difficulties affecting communication, reciprocal social interaction and restricted/stereotyped patterns of behaviour, activities and interests
- Aetiology of ASD is unknown, although it is likely to be multifactorial. It is highly heritable with environmental factors playing a role to a lesser extent
- Diagnosis can be made accurately in the pre-school period
- Early recognition of the condition can have a positive impact on outcome
- Co-morbid conditions are common and should be recognized and managed appropriately
- Yield from medical investigations is low and should be guided by clinical presentation and features of the individual patient

Behavioural eating disorders

Dasha Nicholls

Caro Grindrod

Abstract

Eating disorders are serious mental health disorders characterized by morbid preoccupation with weight and shape, manifest through distorted or chaotic eating. Determined food avoidance in the absence of these cognitions is of uncertain nosological status. Anorexia nervosa (AN), bulimia nervosa (BN) and partial syndromes are relatively common, and early intervention is advisable. Aetiology is multifactorial, with high heritability. Prognosis overall is good but treatment can be long and intensive, significantly impacting families. Essential aspects of management are an integrated multidisciplinary approach, working collaboratively with families and young people when possible. Psychological interventions focus on the eating disorder, supported by medical monitoring and dietetic guidance. Although working with families is the backbone of treatment for AN, young people also need confidential individual appointments. The role of inpatient treatment is evolving. For BN, family or individual approaches may be equally effective. Paediatric expertise is of particular value in the assessment and management of acute malnutrition and complications secondary to disordered eating behaviours, in the early stages of re-feeding, and in the monitoring and management of long-term complications such as growth retardation, pubertal delay and osteopenia.

Keywords adolescent; anorexia nervosa; bulimia nervosa; child; eating disorders

Introduction

Anorexia nervosa (AN) and bulimia nervosa (BN) are characterized by morbid preoccupation with weight and shape, manifest through distorted or chaotic eating (Tables 1 and 2). This behaviour differentiates these disorders from other psychological problems associated with abnormal eating.

Eating disorders are distinct from feeding disorders. The latter term is used for younger children or when feeding skills have

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Diagnostic features of anorexia nervosa (adapted from DSM-IV and ICD-10 criteria)

- Weight lost or maintained at less than 85% of expected weight for height and age, or failure to make weight gain during a growth period
- Fear of gaining weight or becoming fat, even though underweight
- Disturbance in the way one's body weight and shape is experienced (body image distortion), undue influence of body weight or shape on self-evaluation, or denial of the seriousness of low body weight
- Amenorrhoea, i.e. absence of at least three consecutive cycles or delayed pubertal development
- Weight loss is achieved by restriction of food intake and specific avoidance of 'fattening foods' and one or more of the following:
 - Self-induced vomiting
 - Self-induced purging
 - Excessive exercise
 - Use of appetite suppressants and/or diuretics
- If bingeing or purging behaviours are *absent*, this is known as restrictive anorexia nervosa; if *present*, as binge purge anorexia

Table 1

never fully developed. Feeding problems are relatively common and generally resolve with no effects on development. At the more severe end, some continue to have difficulties, affecting growth and development and necessitating specific treatment. Feeding disorders are more common in children with medical or developmental problems, such as significant prematurity, but can occur separately. Eating problems that develop after a period of normal feeding are more likely to be related to underlying psychological issues, as children get older and develop cognitively.

Feeding disorders aside, there is debate about the nature and classification of psychological eating difficulties not related to weight and shape concerns. The Diagnostic and Statistical Manual for Mental Disorder (DSM-IV) contains only three eating disorder diagnoses: AN, BN and eating disorder not otherwise specified (EDNOS). EDNOS is only used for patients who do not fulfil full diagnostic criteria for AN or BN but in whom the characteristic eating disorder psychopathology of weight and shape concern is present, and in whom the eating disorder is clinically significant. The majority of patients of all ages presenting clinically and in epidemiological studies of eating disorders would be classified with EDNOS. The International Classification of Diseases (ICD-10) has a broader concept of eating disorders. Patients with EDNOS are termed 'atypical', but in addition there are diagnoses for 'overeating associated with other psychological disturbances', 'vomiting associated with other psychological disturbances', 'other eating disorders' and 'eating disorder, unspecified', allowing a wider number of clinical presentations, including psychogenic loss of appetite, to be classified. The lack of consensus about the nature of eating problems not associated with weight and shape concerns, such as selective eating, food avoidance emotional disorder and food phobias, means that little

Diagnostic features of bulimia nervosa (adapted from DSM-IV and ICD-10 criteria)

- Persistent preoccupation with eating and recurrent episodes (over a period of months) of binge eating, which are characterized by:
 - Eating a large amount of food in a short period of time
 - A sense of lack of control while eating
- Attempts to counteract the 'fattening' effects of food by use of compensatory behaviours such as:
 - Self-induced vomiting
 - Purgative abuse, alternating periods of starvation or excessive exercise
 - Use of drugs such as appetite suppressants, diuretics, thyroid preparations or, in diabetics, misuse of insulin
- Psychopathology consisting of a morbid dread of fatness and setting of a target weight way below what might be considered healthy
- Bulimia nervosa may follow on from a period of anorexia nervosa, but would only be diagnosed if the patient is no longer significantly underweight

Table 2

has been published to inform clinical practice. For a review, see Nicholls and Jaffa.¹

Apart from recognition and diagnosis, the main areas where paediatric expertise is vital are in the management of malnutrition and other acute medical complications, and of long-term complications such as the impact on growth, development and bone density.

Epidemiology

AN occurs in children as young as 7 years, while BN tends to occur later, from around 12 years of age, and rarely before puberty. The prevalence of AN is around 0.3–0.5%, with a peak age of onset between 15 and 18 years of age. Higher prevalence rates are found in specific high-risk populations, such as athletes, models and ballet dancers. The prevalence of BN is slightly higher, at just under 1%, with a slightly later mean age of onset, but BN is much less likely to come to clinical attention. The prevalence of EDNOS is over three times that of full syndrome AN or BN combined.² The prevalence of AN rose up to the 1970s, possibly due to increased recognition, but since then has remained relatively stable, while the prevalence of BN has been more subject to fluctuation.³ It is a matter of debate whether eating disorders are becoming more common in children and adolescents, although there is some suggestion of this. More cases may be presenting for treatment, due to public awareness, leading to increased healthcare expenditure and professional awareness.

Female-to-male ratios of AN are around 11:1, with slightly more boys in prepubertal samples, and for BN around 30:1. Presentation in boys is similar to that of girls, except that age of onset in boys is later on average, possibly related to the later onset of puberty and its role in the development of eating disorders. The problem of what is, and what is not, classified an

eating disorder may account for some of cultural variation seen in the prevalence of eating disorders.

Pathology and pathogenesis

Biological, psychological and sociocultural factors all have a role in the aetiology of eating disorders, which are often thought of in terms of predisposing factors (risks), precipitating factors (triggers) and perpetuating (maintaining) factors. Clinically, focusing on maintaining factors and ways of overcoming these may be most useful. For example, if an episode of weight-related teasing is identified as a trigger, addressing the bullying will not in itself address the eating disorder.

Understanding of the aetiology of eating disorders has been subject to definite 'fashions'. Recently, there has been increased interest in the neurobiological aspects, thanks largely to advances in neuroimaging, and molecular genetics. This is not to devalue sociocultural theories, but rather to understand better why, within one family, one member but not another is affected. Over 30 risk factors have been identified, suggesting that none is either necessary or sufficient on its own.

The importance of heritance in the development of eating disorders comes from family studies, twin studies and adoption studies, with heritability estimates around 60–75% for AN and 30–80% for BN. There is also considerable evidence for genetic contributions to individual symptoms, attitudes and behaviours, such as self-induced vomiting, or perfectionism traits, which increase risk within individuals. Puberty may also activate some aspect of genetic heritability.

Course of the disease(s)

Eating disorders are often thought to be rooted in difficulties in emotional regulation and expression, the route to recovery being the acquisition of age-appropriate ways to identify and express difficult negative emotions, especially anxiety and anger or protest.

Lask and Bryant-Waugh⁴ describe three stages of recovery during the course of early-onset AN. In Stage 1 the eating disorder is the predominant feature, characterized by preoccupation with food, and weight and shape concerns. At this stage denial is common. Stage 2 brings increased assertiveness, the young person expressing powerful, negative feelings, often directed towards parents or professionals. It is important to warn parents to expect this stage, which can last around 6 months, and to herald it as part of recovery. Thereafter, more age-appropriate expression of feelings become apparent (stage 3). This description of the recovery process fits neatly with the three stages of treatment described in the therapy process.⁵

BN tends to have a more chronic and fluctuating course. Identification is often delayed, the nature of the disorder being easier to conceal than AN. Often conceptualized as a coping strategy at difficult times, and highly mood related, it is often likened to addictive disorders.

Outcome and prognosis

Overall, outcome studies reveal good outcome in adolescents with AN of between 49% and 75% after 10 or more years follow-up,

although even with intensive treatment, recovery can be slow. In adolescents in treatment for AN, Gowers et al⁶ found that fewer than one in five fully had recovered by 1 year, and only one-third recovered by 2 years. For patients who have required hospitalization, an average of 7 years to recovery is often quoted.⁷ For around 10% there is a poor outcome, with the highest mortality and morbidity of any psychiatric disorder.

There are fewer studies on the outcome of adolescent BN, although full recovery is expected in over 50% of patients. EDNOS can be just as clinically severe as AN and BN and is more likely to present with comorbid conditions. AN precedes BN or EDNOS more often than BN precedes AN. Fluctuation in clinical features developmentally and over time have led some to question the validity of distinguishing AN from BN, but rather to take a 'transdiagnostic' approach.⁸ Some genetic and outcome findings, however, would suggest the distinction is valid.

Once the young person has recovered from the eating disorder, secondary psychopathology may remain, most commonly depression or anxiety disorders.

Diagnosis

Diagnosis relies on good clinical interviewing skills, the presence of eating disorder psychopathology being ultimately identified through subjective report by the young person, interviewed alone. Age-appropriate semi-structured interviews are the gold standard, although interview by an experienced clinician is adequate. In younger children, developmental issues related to cognitive level and capacity for self-evaluation⁹ can make it difficult to be certain about the diagnosis based on the young person's account alone, and parent report is necessary. When there is determined food avoidance in the absence of specific eating disorder or other psychopathology, the term food avoidance emotional disorder has been used. This is not a formal eating disorder diagnosis as widely understood. If the diagnosis remains uncertain, starting to increase food intake often clarifies the diagnosis. If fear of weight gain is present, active weight loss behaviours will ensue. Similarly, a fear of swallowing will become more obvious if certain foods are avoided or only liquids can be managed.

Some simple questions are useful to ask as an initial screen for an eating disorder, for example – 'do you think you have an eating problem?' or 'do you worry excessively about your weight?'.¹⁰

The history is best taken with the whole family together, informing them of the need for individual time with the young person, and a physical examination. Choice should be given about when this is done and who the young person would like present. Assessment should be regarded as the first step in treatment and an important opportunity to engage and motivate the young person and family. This translates to avoidance of challenging and confrontation, but to listening and the provision of clear information and guidance. Young people repeatedly say that being listened to and talked to with respect is the most important aspect of the assessment.

Listening to their parents' account of the eating difficulties and the context in which they arose can be helpful to young people, even if they cannot directly contribute to the story – it can help them understand the factors that may have been beyond their control. Ask about current eating patterns and a typical day's intake, as well as specific questions about compensatory

behaviours. Current intake gives important information regarding risk of nutritional deficiencies, and is important in establishing how the re-feeding should be tailored safely. Onset is often slow and insidious, with acute deterioration once it comes to light. Children often stop drinking as well as eating.

A full history also addresses family history of mental disorder, and family relationships, including the role of the extended family, identifying areas of tension and potential support. Marital relationships are only of relevance in relation to parents' capacity to work together in the interests of their child. Family attitudes and beliefs about food, weight and shape influence the way that eating disorders are addressed within the family context and may therefore affect prognosis. Social context (housing, employment and financial situation) and practical considerations are important for treatment planning. A developmental history should include feeding and early attachment, and premorbid personality including perfectionism, peer relations, obsessional traits, separation anxiety, autism spectrum disorder traits and self-esteem.

Eating disorder psychopathology, mood and psychological risk (e.g. suicidal ideas or self-harm) are best assessed individually. Ask about eating patterns and current intake, dietary restrictions and rules (such as set calorie limits, eating at certain times), compensatory behaviours (purging, laxatives, exercise) and binge eating. Beliefs about weight and shape, preoccupation with weight and shape, concerns about eating, fear of weight gain, self-evaluation with respect to weight shape or eating and motivation to change are all key to making a diagnosis and treatment plan.

Adequate time should be allowed for feedback and decision-making at the end of the first assessment, given frequently appropriate anxiety about the need for change.

Physical examination (Table 3)

AN carries considerable serious physical risks and needs careful monitoring. In BN, physical problems are caused by frequent vomiting and potential excessive use of laxatives. Some of the complications of AN and BN are due to lack of energy, some to metabolic disturbance and some to endocrine disturbance (of hypothalamic origin) (Table 4). Some are potentially life-threatening, whilst others are associated with long-term compromise of health. Complications that are unique to younger patients are growth retardation, pubertal delay or arrest, and reduction of peak bone mass. An atypical picture needs an open mind and a thorough medical review. Common differential diagnoses include gastrointestinal disease such as Crohn disease, chronic disease affecting appetite and growth such as renal failure, endocrine disorders, intracranial pathology and other psychiatric disorders such as obsessive-compulsive disorder and depression.

Nutritional assessment should consider the past, the present and the future: duration of low weight, rapidity of weight loss, menarcheal status, body mass index (BMI) centile (or % median BMI), haemodynamic stability and future predicted intake (more commonly over- than under-estimated). Fluid intake may be restricted (to lose weight) or excessive (to increase weight temporarily). Rapid weight loss (more than 1 kg/week) can cause medical instability even if the child is not underweight. Muscle weakness and peripheral neuropathy are signs of serious nutritional deficit. Local protocols agreeing thresholds for paediatric admission can be helpful.

Physical examination in eating disorders

What to look for on physical examination	When to worry
Weight, height and BMI centiles (or % median BMI if below 2nd centile) falling or below 9th BMI centile	<85% BMI for age (between 2nd and 9th centile) is underweight. <75% BMI, serious/consider admission
Bradycardia and orthostatic changes in pulse or blood pressure, based upon age-appropriate norms	Pulse <50 (45 at night); BP <80/50; orthostatic changes in pulse (>20 bpm) or blood pressure (>10 mmHg)
Hypothermia	<35 °C
Dull, thinning hair	
Sunken cheeks, sallow skin/skin integrity	
Lanugo hair	
Delayed pubertal development for age/atrophic breasts	No signs of puberty at 13; premenarcheal at 15
Pitting oedema in peripheries	
Cold extremities/acrocyanosis/weak peripheral pulses	Normal capillary refill <1 s
Dehydration (skin turgor, mouth, tongue)	If visible in older child, suggests >5%
Muscle wasting	Difficulty sitting up from supine, and rising from squat to standing (SUSS test) without use of hands
Signs of bingeing/purging, e.g. dental erosion, callouses on fingers	
Signs of vitamin deficiency	

Table 3

Growth slows down and even stops during a period of starvation. After starvation is over, catch-up growth can occur but it is still unclear to what extent. Our best guess for the 'dose' of starvation needed to have a permanent effect on height is 4 years before completion of growth. There are case reports of people going through puberty in their mid to late 20s, and anecdotal accounts of menarche at nearly 50.

Between 25% and 40% of young people with AN will have osteopenia on bone density scan. The long-term fracture risk is around three times that of the general population. Interpretation of reduced bone density in AN in young people should consider the impact of pubertal delay and growth failure on bone size.

Management

The treatment of eating disorders in children and adolescents presents many challenges to the clinician. Young people with AN are terrified at the thought of eating and weight gain, and at best ambivalent about receiving help. They may be suffering physical

Medical complications of calorie restriction and purging

Calorie restriction

- Cardiovascular
 - ECG abnormalities – bradycardia
 - T-wave inversion
 - ST segment depression
 - Prolonged QT interval
 - Dysrhythmias (SVT, VT)
 - Pericardial infusions
- Gastrointestinal system
 - Delayed gastric emptying
 - Slowed gastrointestinal motility
 - Constipation
 - Bloating
 - Fullness
 - Hypercholesterolemia
 - Abnormal liver function (carotenemia)
- Renal
 - Increased blood urea (from dehydration and reduced GFR) with increased risk of renal stones
 - Polyuria (from abnormal ADH secretion)
 - Depletion of Na and K stores
 - Peripheral oedema with re-feeding due to increased renal sensitivity to aldosterone
- Haematology
 - Leucopenia
 - Anaemia
 - Iron deficiency
 - Thrombocytopenia
- Endocrine
 - Sick thyroid syndrome (low T3)
 - Amenorrhoea
 - Growth failure
 - Osteopenia
- Neurological
 - Cortical atrophy
 - seizures

Medical complications of Purging

- Fluid and electrolyte imbalance
 - Low K
 - Low Na
 - Low Cl
- Chronic vomiting
 - Oesophagitis
 - Dental erosions
 - Oesophageal tears
 - Rarely rupture and pneumonia
- Use of ipecac/laxatives
 - Myocardial damage
 - Renal stones
 - Low Ca
 - Low Mg
 - Low KCO₃
- Amenorrhoea

Table 4

effects from their eating behaviours, impairing their capacity to think. Many patients do not accept that they are unwell, and are often brought to treatment by family members.

Parents often experience first-line healthcare professionals as minimizing eating difficulties on initial presentation.¹¹ It is crucial that weight is monitored in a standardized way as soon as concerns arise. Eating disorders are unlikely to resolve on their own, so a 'wait and see' approach is contraindicated. Ongoing weight loss should alert concern, but parents often report changes in behaviour, such as social withdrawal, altered eating behaviours, secretiveness, and ritualized and restricted activities, long before low weight is apparent.

Written information given early in treatment is important, followed by the time to answer questions and to discuss areas of concern. Professionals should recommend resources for parents and children on eating disorders in the younger population (see below). Parents and children need a clear statement about diagnosis, the likely course and possible complications of the illness, and proposed treatment.

The aims of treatment are to reduce risk, encourage weight gain and healthy eating, reduce other related symptoms, and facilitate psychological and physical recovery.

The evidence base for effective treatments is limited, and existing guidelines, such as the NICE guidelines,¹⁰ are largely based on consensus expert views. Treatment must address both physical and psychological aspects of the condition. Early intervention in a developmentally appropriate treatment setting is likely to produce the best outcome. For more established illness, the aim is to live with the disorder in a way that does not compromise health. A combination of integrated interventions offered by a multidisciplinary team is needed. In most situations, parents should be involved in treatment.

Currently, medication has only a small part to play in the management of eating disorders. In AN, depression often lifts with improved nutritional state. If not, antidepressants, usually selective serotonin re-uptake inhibitors (SSRIs) such as fluoxetine, may have value. Medication should be used with caution due to the increased risk of underweight, which must be clearly explained to the patient and family. Evidence for the effectiveness of SSRIs in BN is stronger, with high-dose fluoxetine being the treatment of choice, although there are no studies exclusively in adolescents.

Occasionally nutritional supplements and hormonal support are indicated, most often as damage limitation in chronic illness. These should only be considered by specialists and in conjunction with the rest of the treating team. In particular, the use of hormone replacement for the treatment of impaired bone density is rarely indicated. The mainstay of treatment remains weight gain and nutritional rehabilitation. Improvement in bone density is not usually seen in the first year of treatment, but after 1–2 years.

Re-feeding

The aim of re-feeding is healthy weight restoration in the least invasive way. Wherever possible, re-feeding is done orally and in the home if safe. The child needs clear expectations about what they need to manage, and dietetic input can be very helpful with this. The aim for weight gain is 0.5 kg for outpatients and between 0.5 and 1 kg for inpatients. This generally requires between 3500 and 7000 extra calories a week. Starting with low intake and building up slowly is safest.

Psychological interventions

In the community

Ideally, young people with eating disorders should be treated as outpatients, incorporating psychological and physical treatment, by healthcare professionals competent to give this treatment and in assessing physical risk. Although working with families is the backbone of child and adolescent mental health, children and adolescents should also be offered individual appointments separate from their family or carers. Psychological interventions need to focus on both the eating behaviours and the young person's thoughts about their weight and shape, alongside clear expectations for weight gain in the case of AN. Effective treatment requires a skilled multidisciplinary team who are able to work collaboratively, in a good alliance with the family.

For AN, the first-line treatment is family therapy, weekly or more frequently at first, supported by regular medical monitoring and dietetic input. The model for which there is the greatest evidence supports parents being in charge of their child's eating until the young person is well enough to share responsibility. Family interventions both address and focus directly on the eating disorder. This work can be undertaken with the family all together (conjoint family therapy) or with parents separately (parental counselling). If separate, it is usual to offer individual therapy to the young person too. Individual therapy has a role, especially for older adolescents who might be expected to take more responsibility, although the evidence base is weaker. If obsessive-compulsive features are marked, progress in treatment may be slower.¹²

Treatment trials for adolescents with BN were largely absent until recently, but two recent studies merit mention. In the first, Schmidt et al¹³ found that guided self-care, using a cognitive behavioural model, was equally and more rapidly effective at reducing eating disordered behaviour than family therapy, and was more cost-effective and acceptable to the young people. By contrast, Le Grange et al¹⁴ found that family therapy was both more effective and more rapid than an individual-based treatment, in this case supportive psychotherapy. In practice therefore, it seems reasonable to offer choice and assess the individual circumstances of the young person and their willingness to involve, and likely support of, their parents.

Alongside outpatient treatment, monitoring of growth is required, especially for pre-menarcheal children. Regular progress reviews are important and all professionals involved should have clear, documented roles and responsibilities.

Increasingly, more intensive methods of outpatient treatment are being sought, because of questions about the cost-effectiveness of inpatient treatment. Outreach services can contribute to decreased need for inpatient admission, and intensive family approaches such as 'multifamily therapy' are undergoing trials.

In hospital

The decision to admit a young person with an eating disorder is made for one of four reasons:

1. A rapid deterioration in medical state;
2. Marked depression, suicidal ideation or intent;
3. Other major psychiatric disturbance;
4. Failed outpatient treatment.

Care may be on a paediatric ward or a child or adolescent psychiatric inpatient ward. Paediatric admission works best if close links remain with other elements of treatment and if locally agreed protocols are in place between paediatric and mental health services. It is helpful to distinguish the need for medical stabilization from re-feeding. Young people may find it easier to eat at home, if the risks can be managed.

Psychiatric admission is an altogether more serious decision, admissions usually being for 4–6 months or more. The following need to be considered prior to psychiatric admission for an eating disorder:

- Care should be provided in an age-appropriate setting.
- Treatment should be provided as close to home as is possible.
- The potential side effects of inpatient admission need to be considered, including isolation from family and increased resistance, and needs to be balanced with the educational and social needs of children and adolescents.
- The long-term effects on the family, in terms of time commitment and emotional effects of admission.
- A plan needs to be in place to aid transition to local outpatient services on discharge.

There is a limited role for inpatient services in the care of patients with BN, except in the management of suicide risk and self-harm.

Psychiatric admission usually follows failure of outpatient treatment. However, a recent randomized controlled trial⁶ found no advantage in outcome of inpatient over outpatient psychiatric care, and as such ‘challenge the intuitive clinical belief that a step up from outpatient to inpatient psychiatric care is indicated for those who fail to make progress’.⁶

Weight gain of 0.5–1 kg/week is usual in underweight patients, with familiarity of safe re-feeding practice needed in the initial stages. Once in a healthy weight range, meal plans will need to be adapted to allow for continuing growth and the nutritional demands of puberty. Although weight gain is ultimately associated with improvement in all aspects of functioning, initially it may increase eating disordered behaviour (in an attempt to eliminate the extra weight), anxiety and distress. Adequate weight gain may reduce the risk of readmission, whilst the speed should be fast enough to avoid negative institutionalization effects but not so fast as to cause overwhelming anxiety or risk from re-feeding. Nasogastric feeding is considered only when patients are medically compromised or are unable to gain weight with supported meals and behavioural re-feeding regimens.

Consent to treatment is needed and treatment against the patient’s or parents’ wishes is always a last resort. Treatment against consent is a highly specialized procedure requiring expertise in the care of patients with severe eating disorders. It can be done in the context of the Mental Health Act 1984 or Children’s Act 1989, which allow a young person’s refusal of treatment to be over-ridden. Parental consent should not be relied on indefinitely, and clinicians should ensure the legal basis for clinical action is clear. NICE recommends seeking a second opinion when consent issues are highlighted.¹⁰

Adolescents’ views of treatment

Young people with eating disorders say that without the motivation to get well, they struggle to make use of treatment. They value being listened to, and their views respected, even if

ultimately decisions need to be made against their wishes. When treatment is experienced as disempowering or punitive, they tended to reject and fight against it.

Parents’ views

What parents find helpful includes ‘being firm and presenting a united front, support and understanding, connecting with other parents in similar situations’.¹¹ Parent peer support groups are very powerful in this respect. Structured interventions and a skills manual for carers of people with an eating disorder have been developed.¹⁵ Parents want clinicians to include them in treatment, support and guide them in their child’s care and demonstrate positive attitudes toward them. The implications for clinicians include the need for sensitivity to parents’ vulnerability, ensuring congruence between clinicians’ and parents’ expectations about treatment, and strengthening formal channels of communication. Around 50% of carers suffer anxiety and around 13% will have significant depression.¹⁶

Follow-up

Length of follow-up is best determined by how long the young person and family need support. It is important not to focus solely on whether eating disorder symptoms are present, but to think more broadly about the extent to which coping strategies for the future stresses have been developed, ongoing risk and impairment, and whether the young person is equipped for developmental tasks appropriate to their age. In practice, young people usually want to leave outpatient treatment before clinicians and their parents want to discharge them. Regular reviews of progress and treatment are needed to inform changes in treatment intensity. In general, outpatient treatment for a minimum of a year after an inpatient admission is advisable. Monitoring of physical outcomes, e.g. bone density and menstruation, may need to continue beyond psychological intervention. Careful transition to adult services is needed for chronic cases.

Prevention

The efficacy of prevention is not yet established, although there have been some promising inroads. Targeting mental health and self-esteem seems to be more effective than targeting weight and eating behaviours. There are concerns about the impact of antiobesity messages such as ‘fat is bad’ for children with a perfectionist and literal mind. ◆

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- www.nice.org.uk/nicemedia/pdf/cg009niceguidance.pdf NICE guidelines for eating disorders.
- www.rcpsych.ac.uk/files/pdfversion/cr130.pdf Nutritional guidelines for the management of anorexia nervosa.

Practice points

- Diagnosis should ideally not be based on either a parent's or young person's report alone
- Eating disorders are rarely self-limiting and, when suspected, should be monitored carefully
- The most important skill is engagement, for which a collaborative and information sharing stance is most helpful
- Most patients should be treated on an outpatient basis, provided risks can be managed safely, but both physical and psychological risks can increase at first with intervention
- The burden of caring for a young person with an eating disorder is enormous and adequate support for parents and siblings is essential

Obsessive–compulsive disorder in children and adolescents

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Abstract

Despite obsessive–compulsive disorder (OCD) being one of the more common serious mental illnesses, it continues to be shrouded in shame and secrecy. The shame surrounding the condition, in combination with a lack of recognition of its defining symptoms, can lead to delays of several years before it is diagnosed. Children with OCD frequently present to non-psychiatrists for treatment. It is important that paediatricians familiarize themselves with the characteristic symptoms. This review summarizes current research regarding the epidemiology and aetiology of OCD, the assessment of childhood-onset OCD, as well as its psychological and pharmacological management.

Keywords adolescents; assessment; children; cognitive behaviour therapy (CBT); obsessive–compulsive disorder (OCD); selective serotonin reuptake inhibitors (SSRI); treatment

Introduction

Earlier studies reported obsessive–compulsive disorder (OCD) in 1–2% of children; more recent estimates of prevalence are 0.25% in 5–15-year olds. OCD can occur throughout the lifespan. It may occur in children as young as 6 or 7 years old, and most newly diagnosed adults report onset of symptoms in childhood or adolescence.¹ It has an equal incidence in both sexes.

Aetiology

The aetiology of OCD is not well understood, but there has been extensive research on environmental and genetic risk factors. Twin, family, segregation and linkage studies strongly support a genetic component for OCD. Most studies looking at candidate genes have focused on genes in the serotonergic and

dopaminergic pathways, but none has reached genome-wide significance.²

Neuroimaging studies have identified alterations in brain frontostriatal circuitry and in the basal ganglia in patients with OCD. One paediatric study demonstrated a smaller globus pallidus volume in unmedicated patients with OCD compared to healthy controls.³ Recent research in adults suggests that the dorsal prefrontal cortical and bilateral midbrain grey matter abnormalities in OCD appear to be primarily driven by the effects of OCD symptom severity.⁴ Other studies suggest that there may be ‘brain-markers’ for OCD even in the absence of symptoms; for example, a recent study demonstrates orbitofrontal brain dysfunction not only in individuals with OCD, but also in their unaffected close relatives.⁵

Although there is no evidence for life events as a cause of OCD in susceptible individuals, these may trigger relapse or exacerbate symptoms. This could be at the time of school transitions, exams or relationship difficulties with peers or family.

Definitions

In the WHO ICD-10 Classification for Mental and Behavioural Disorders, the following must be present for the full clinical diagnosis for OCD to be made.

- *Either obsessions or compulsions or both present on most days for a period of 2 weeks.*
- *Obsessions* (unwanted ideas, images or impulses that repeatedly enter a person’s mind) and *compulsions* (repetitive stereotyped behaviours or mental acts driven by rules that must be applied rigidly) share the following features:
 - Patient is aware that these originate from their own mind;
 - Repetitive unpleasant and distressing to the patient. At least one is perceived as excessive or unreasonable (‘egodystonic’);
 - At least one is resisted unsuccessfully, even though others may be present which the sufferer no longer resists;
 - Thought of carrying out the obsession or compulsion is not intrinsically pleasurable (simple relief of tension momentarily on completion of the thought/act is not regarded as pleasure in this sense).
- The symptoms must be *disabling*. Even young children will have some insight into the *senselessness* of the thoughts and behaviours.⁶

Typically, the child experiences an escalating vicious cycle of obsessions and compulsions. The obsessions generate significant anxiety, and rituals develop as a way of temporarily reducing the anxiety levels. After ritual completion, anxiety levels soon rise again, further fuelling the obsessions, and so the cycle continues. The temporary relief of anxiety with the compulsions negatively reinforces the behaviours and prevents habituation to the anxiety. Eventually, the rituals become more automatic and increase, rather than reduce, the anxiety, so that no temporary relief is experienced.

These symptoms are therefore easily distinguishable from the ordinary childhood rituals that are common in early childhood. The latter may include bedtime rituals with a parent or routines about food, toys or clothes, but in the absence of other problems they are non-distressing, do not take up excessive time or stop the child from doing other things, and are self-limiting.

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Assessment and diagnosis

There can be a long delay in the diagnosis of OCD due to lack of recognition or, often, because sufferers are embarrassed by their symptoms and may be unaware that they can be helped. The presence of certain symptoms (Table 1) and a simple six-question screen may alert the clinician to the possibility of a diagnosis of OCD (Table 2). A short screening instrument suitable for use in children is also available: the short OCD screener (SOCS). This has a sensitivity of 0.97 (95% CI 0.91–0.98) to detect OCD cases, although further assessment is required to confirm the diagnosis.⁷ If a diagnosis is suspected, then a more thorough assessment can be completed. For more complex cases it is advisable to refer the child to the local Child and Adolescent Mental Health Service (CAMHS). The child and carer(s) are invited to the assessment; it may be helpful to involve siblings. Most CAMHS will typically carry out the following assessment.

- A detailed history of obsessions and compulsions is taken:
 - This in part should be done with the parents giving a history, and perhaps other family members such as siblings.
 - Engagement with the family is essential, as they are likely to be central to successful treatment. The whole family needs to understand the diagnosis and techniques for its management.
 - It is also important to involve the family in the discussion as they can often give information about how the child involves other family members in their compulsions, and how the condition impacts on everyday life, e.g. making everyone late for school because of morning rituals, or limiting activities because of fear of contamination.
 - A helpful technique, referred to as ‘externalizing the problem’, is to acknowledge how the condition makes the child do things, rather than saying that the child makes everyone late for school. This can enable everyone to talk about the symptoms without feeling that the child is being blamed. Quite often, the child may even have a name for their OCD.
 - Younger children may find it difficult to identify the underlying obsession driving a compulsion.
- A detailed early developmental history, family history, and past medical and surgical history (and brief physical examination) should be undertaken to identify the risk factors outlined in Table 1.

Symptoms and risk factors that may alert you to obsessive-compulsive disorder (OCD)

- Anxiety
- Depression
- Tics
- Sydenham chorea and rheumatic fever
- Recent streptococcal infection (ENT)
- Chapped hands, eczema from handwashing
- Trichotillomania
- Hypochondriasis: fear of cancer, fear of dying, fear of HIV
- Concerns about body appearance (body dysmorphic disorder)
- Strong family history of OCD, anxiety, tics, depression

Table 1

Screening questions for obsessive-compulsive disorder recommended by NICE

- 1 Do you wash or clean a lot?
- 2 Do you check things a lot?
- 3 Is there any thought that keeps bothering you that you would like to get rid of but cannot?
- 4 Do your daily activities take a long time to finish? (e.g. getting ready for school)
- 5 Are you concerned about putting things in a special order or are you very upset by mess?
- 6 Do these problems trouble you?

Table 2

- Due to the sensitive nature of the symptoms, the child should also be seen on their own for a mental state examination to assess for OCD and related co-morbidities. The use of the validated CYBOCS (Children’s Yale–Brown Obsessive Compulsive Scale)^{8,9} semi-structured checklist is a useful adjunct to generate a list of current and historical symptoms and to gauge severity. Typically, a child is asked about compulsions first as these are easier to identify.

The most common obsessions include

- fear of causing harm to someone else
 - fear of causing harm to self
 - fear of contamination
 - need for symmetry or exactness
 - sexual and religious obsessions
 - fear of behaving unacceptably
 - fear of making a mistake.
- The most common compulsions include
- Behaviours
 - cleaning
 - handwashing
 - checking
 - ordering and arranging
 - hoarding
 - asking for reassurance.
 - Mental acts
 - counting
 - repeating words silently
 - ‘neutralizing thoughts’.
 - Enquire about symptoms that may identify co-morbid disorders and related OCD spectrum disorders.

Co-morbidity, related disorders and differential diagnoses

During the assessment it is important to identify co-morbidity. OCD co-occurs¹⁰ (Table 3) and overlaps with many disorders, which are sometimes referred to as the obsessive-compulsive spectrum disorders (Table 4).¹¹ Co-morbidity such as oppositional defiant disorder, for example, may make the most dominant condition more difficult to treat and may impact on the treatment strategies used.

For paediatricians, the most likely differential diagnoses are autism spectrum disorders and Tourette syndrome, although

Conditions that commonly co-occur with obsessive-compulsive disorder

- Depression
- Specific phobia
- Social phobia
- Eating disorder
- Alcohol dependence
- Panic disorder
- Tourette syndrome

Table 3

both of these groups may co-occur with OCD. A subgroup of children with OCD may fall into the PANDAS category.

PANDAS

Patients with Sydenham chorea frequently have emotional and/or behavioural symptoms, particularly OCD. Sydenham chorea is associated with group B haemolytic streptococcus infection. In the late 1990s, Swedo's group described 50 patients, who did not have Sydenham chorea, but did develop OCD and/or tics in response to a post-streptococcal autoimmune response. This type of OCD is referred to as PANDAS (paediatric autoimmune neuropsychiatric disorders associated with streptococcal infections). The criteria for PANDAS are¹²

- presence of OCD or tic disorder
- onset between 3 years and beginning of puberty
- abrupt onset of symptoms and a fluctuating course characterized by dramatic exacerbations of symptoms
- onset or exacerbations of symptoms is temporally related to infection with Group A beta haemolytic streptococcus
- abnormal neurological examination (hyperactivity, choreiform movements and/or tics) during an exacerbation.

It was proposed that, through a process of molecular mimicry, autoantibodies were generated against the basal ganglia, leading to behavioural disorder.¹³ There is a body of evidence to support this hypothesis and there is now also evidence to suggest that some children with 'typical' OCD may have antibasal ganglia antibodies. Central nervous system autoimmunity may therefore have a role in a subgroup of cases of OCD.¹⁴

Autism spectrum disorders/pervasive developmental disorders

The repetitive and stereotyped patterns of behaviour in autism can resemble some of the compulsive symptoms of OCD. A

major difference between the two disorders is the underlying emotion that is associated with the rituals. A diagnosis of OCD, by definition, requires an element of internal anxiety and distress experienced if the obsession/compulsion is interfered with, and the child with OCD has insight into the unnecessary, time-consuming and unwanted nature of the rituals. In contrast, the child with autism enjoys the rituals and sees no reason to try to limit or stop them.¹⁵

The rituals typically seen in autism are part of the third symptom domain of autism: restricted, stereotypical, repetitive repertoire of interests and behaviours, and are in the context of autistic rigidity. The autistic child may insist on the performance of particular routines and rituals and have stereotyped, often unusual, preoccupations. Quite often, disruption of these routines can lead to a significant behavioural disturbance.

Children with autism are at a significantly increased risk of psychiatric co-morbidity (this risk is further increased in the presence of mental retardation). Children with autism may therefore have co-morbid OCD, but frequently clinicians attribute all repetitive phenomenology to the autism, often because it is difficult to differentiate the two syndromes.¹⁶ Where appropriate, co-morbid OCD should be detected and treated if present. Co-morbidity of OCD in children with autism can be difficult to demonstrate for a number of reasons.

- It is often difficult to assess the mental state of children on the autism spectrum.
- Many children with autism have associated mental retardation and are non-verbal; hence it is difficult to infer an internal state. This can only be done through behavioural measures (reduced sleep, reduced appetite, pulse rate increase, sweating, pacing, agitation, papillary dilatation, cold and clammy).
- The anxiety caused by stopping a compulsion could easily be confused with the distress caused by the stopping an autistic ritual (because of the cognitive rigidity of the autistic child).

In addition, motor tics, OCD and affective disorders are common in relatives of autistic probands.¹⁷

Co-morbid autism should also be looked for in those diagnosed with OCD,¹⁸ especially in those who are treatment resistant. A diagnosis of OCD is perhaps more easily made in high functioning autistic children where they are able to verbalize their internal distress regarding their obsessions and compulsions, and are able to convey that they have some notion of the senselessness.

Treatment

In 2004, a multicentre randomized controlled trial compared cognitive behaviour therapy (CBT), sertraline [a selective serotonin reuptake inhibitor (SSRI)] and a combination of these in children and adolescents, and concluded that children with OCD should begin treatment with CBT alone, or CBT plus an SSRI.¹⁹ In 2006, NICE published the evidence-based treatment options for OCD (and body dysmorphic disorder) for young people and adults, following a stepped care treatment model²⁰ (Figure 1). Psychoeducation and CBT were recommended as first-line treatment, and then CBT combined with an SSRI for more severe cases. A Cochrane review in 2006 concluded that, although the evidence base is small for psychological treatment, clinical consensus recommends the use of CBT, which can lead to better outcomes when combined with medication compared to medication alone.²¹

Conditions related to obsessive compulsive disorder

- OCD spectrum disorders
- Body dysmorphic disorder
- Trichotillomania
- Hypochondriasis
- Tourette syndrome
- Compulsive hoarding

Table 4

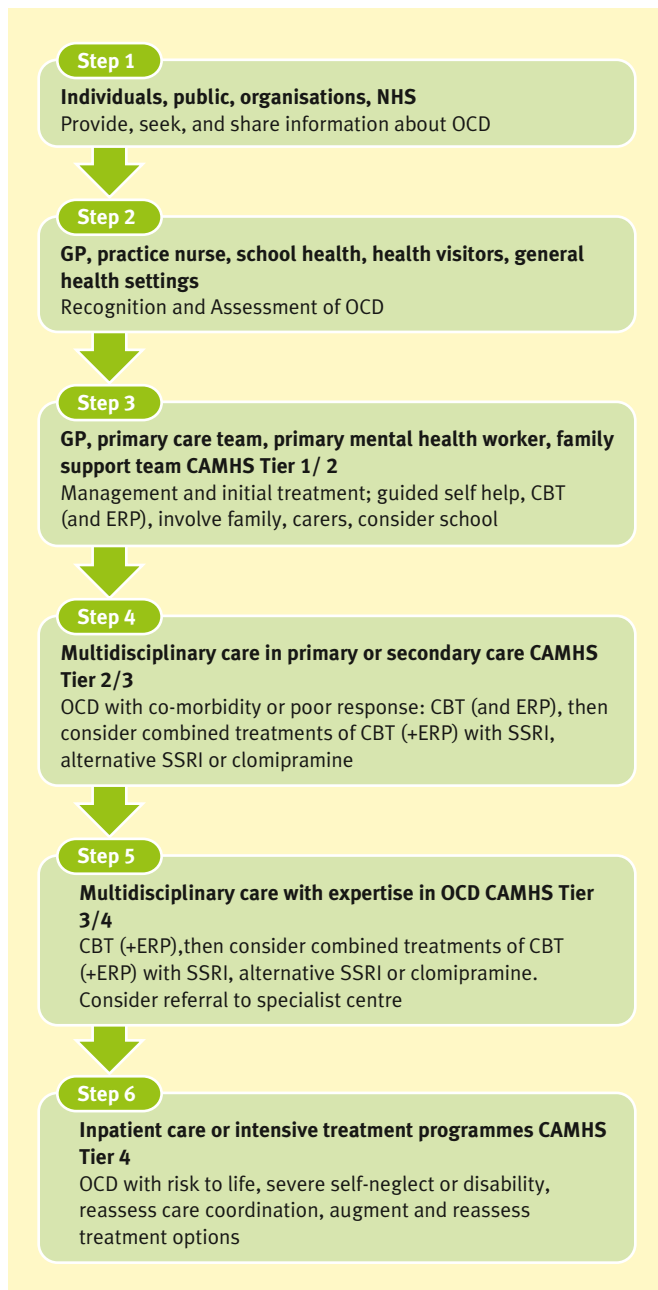


Figure 1 Stepped care model for treatment of obsessive-compulsive disorder (OCD) in children.

Psychological treatment

Mild functional impairment

Very early-onset or mild OCD should be managed with psychoeducation and guided self-help. A list of additional resources is given at the end of this review. Psychoeducation should include an explanation of what obsessions and compulsions are, and the mechanisms of anxiety, as well as the principle of learning to 'face the fear' and cut back on rituals gradually.

Moderate or severe functional impairment

This should be managed with CBT with exposure and response prevention (ERP) and for younger children usually involves the family.²² By involving parents as co-therapists, they can play a

key role in shaping treatment, and encourage and reward the child for progress. It also shows parents how they may be inadvertently reinforcing OCD behaviours, and helps them learn to tolerate their own distress when aiding their children in ERP tasks.

In CBT (usually a brief therapy of 12–14 sessions), patients work collaboratively with the therapist to learn gradually to confront their feared stimuli (e.g. perceived contamination); this is *exposure*. In conjunction with this, they learn to understand and tolerate graded amounts of anxiety while resisting carrying out compulsions; this is *response prevention*. The patient works through a hierarchy of feared stimuli, starting with the least feared of the stimuli, and practices facing the situation whilst monitoring their anxiety levels, observing that the anxiety levels subside eventually without the need for a ritual. These 'experiments', exposure and response prevention, need to be practised several times a day, as over time a patient will have avoided many feared stimuli or developed rituals. CBT with ERP has response rates of 40–85% for those who complete treatment.²³

There is no evidence to support the efficacy of psychodynamic psychotherapy in OCD and NICE does not recommend its use.

Medication

For children with more severe OCD who have shown limited response to CBT incorporating ERP, there is evidence supporting their treatment with SSRIs for symptom remission and improvements in global functioning. These can be used in combination with CBT. At the time the NICE guidelines were published, 18 published trials were identified, of which four did not meet the inclusion criteria. The 14 included studies provided efficacy data from 1034 participants and tolerability data from 1068 participants.

Sertraline and fluvoxamine are licensed for use in children in the UK, but other SSRIs may be chosen for clinical reasons. Sertraline is licensed from the age of 6 and fluvoxamine from the age of 8 years.²⁰ In practice many clinicians find that fluvoxamine produces unacceptable gastrointestinal side effects, including nausea and vomiting.

Fluoxetine is the only recommended SSRI for use in the treatment of depression in children under 18 years old. For children with OCD co-morbid with depression, this is the preferred first-line medication.

Patients with OCD require a trial of an SSRI for at least 12 weeks at the maximum tolerated therapeutic dose. The dose should be increased gradually, frequently with no therapeutic response noticeable on a low dose or for 4 weeks. It may take several weeks to build up to the therapeutic dose, and then the full effects of this may not be evident for several more weeks. If a trial of one SSRI fails, the patient should be reassessed, clarifying compliance and ensuring that co-morbidity has not been missed. A trial of a different SSRI or clomipramine, a serotonergic tricyclic antidepressant, should be considered.

Nausea and appetite loss in the first few weeks, before there is a therapeutic response, may be seen, but is usually tolerated and resolves. SSRIs are also associated with behavioural activation syndromes, which have been linked to a small increase in the relative risk of self-harm and suicidal thoughts (but not completed suicide). Monitoring is especially important in the early stages of treatment and at times of change in dosage. SSRIs should not be

used if a child has renal or hepatic problems, and occasionally they have been associated with increased seizure frequency.

As for all medications in children, the potential risks of untreated OCD, including a potentially life-long impact on emotional wellbeing, social and educational development, need to be considered against the risks of medication side effects, both acute and long term.

Medication may also be indicated in those whose capacity to access CBT is limited by learning disabilities, although every attempt should be made to modify CBT protocols for the less able child. Smaller doses and doses increasing over a longer period of time are recommended for children with learning disabilities, as they have a tendency to develop side effects more quickly.

Medication has occasionally been used where there is no availability of CBT. It is not acceptable to use medication instead of CBT unless the child is unable or unwilling to use psychological treatment. Whilst medication should not be withheld from the child who needs treatment for their OCD, parents and clinicians should be aware of the NICE recommendations that all young people with OCD should be offered CBT; if necessary, the child should be referred to a specialist who can provide CBT or the primary care trust children's commissioner should be asked to change local provision.

Patients refractory to treatment

Some children with OCD may fail to respond to an initial SSRI administered for at least 12 weeks at the maximum tolerated dose, in combination with an effective trial of CBT incorporating ERP. These children should usually have additional trials of at least one other SSRI. Following this, if response is still limited, the child should be referred to a specialist centre. Often children with more severe or chronic OCD have co-morbidities that can affect the initial response to treatment and long-term prognosis.

Conclusions

Specialist child and adolescent psychiatry provision is still very variable throughout the UK, although there is now a nationally commissioned service for the most severe, treatment-refractory OCD.²⁴ Given the hidden nature of OCD, it can take many years for children to be seen in specialist child mental health services. Paediatricians as part of their routine history taking should enquire about a child's worries and mood, and where possible have the opportunity to talk to older children without their family present. Asking an anxious or worried child a few specific questions about OCD symptoms may allow them to reveal their symptoms for the first time, and lead to diagnosis and effective treatment.

At service level, primary care trusts should ensure specialist CBT is available as it is the NICE treatment of choice for a number of mental health issues in children, including OCD. ◆

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FURTHER READING AND USEFUL RESOURCES FOR CHILDREN AND FAMILIES²⁵

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- www.ocdaction.org.uk. National UK Charity OCD Action.
- www.ocdyouth.info. South London and Maudsley NHS Trust Information on OCD and how to recover.

Practice points

- Obsessive-compulsive disorder (OCD) often presents for the first time in childhood or adolescence
- A clinician should ask specific screening questions if OCD is suspected, as diagnosis is often delayed because the child is embarrassed to mention their rituals or worries
- At least 70% of young people with OCD will respond well to the evidence-based treatments, so detection and referral for treatment is important
- There are NICE Guidelines for the assessment and treatment of OCD; it is recommended that people with OCD should be offered cognitive behaviour therapy incorporating exposure and response prevention
- Some children and adolescents with OCD will benefit from treatment with serotonin reuptake inhibiting medication, which should be initiated by a specialist
- OCD can be relapsing or chronic; people with obsessive-compulsive disorder who relapse should have rapid access to services

Conduct disorders in children and adolescents

Karen Baker

Abstract

Conduct disorders in children and adolescents are common problems presenting to professionals involved with child health. This review describes their prevalence and aetiology, as well as relevant evidenced-based interventions crucial to successful management.

Keywords conduct disorders; review

Introduction

Conduct disorders or related behaviours in children and adolescents are important issues in current times. The excessive presence of non-compliance in a child, with or without aggression, produces enormous challenges to parents and teachers. There is a growing awareness of the financial and social costs to the wider community. Children with conduct problems often falter in their interpersonal relationships and struggle to function in a socially effective way.

Children are born with a remarkable potential for emotional and social development. It takes time for them to learn to behave 'properly', i.e. to the relevant socially and culturally defined norms. Aggressive or oppositional responses are not always in themselves inappropriate behaviour. The key for an individual is in recognizing how, when and where this kind of response may be socially acceptable. One way to understand violent or antisocial behaviour in children and adolescents is as a failure to acquire the most appropriate social response alongside the development of undesirable or ineffective behaviours.

Definitions

Establishing when antisocial behaviours necessitate professional intervention has been a challenge. Conduct difficulties have needed to be distinguished from the 'normal' emergence of oppositional behaviours as part of a growing sense of individualization and autonomy in an age-appropriate way. The 'terrible twos' or 'teenage tearaway' developmental phases seen in many children, while challenging to parents and other adults caring for children, are to some degree normal, important stages in the child's psychosocial development. However, some children's and

adolescent's oppositional or antisocial behaviours are in excess of the ordinary.

Both of the recognized psychiatric classification systems (ICD-10 and DSM-IV) define conduct disorder as a repetitive and persistent pattern of aggressive, defiant or antisocial behaviour. DSM-IV further delineates two categories of antisocial disorder in childhood: oppositional defiant disorder (Table 1) and conduct disorder (Table 2).¹ These categories are recognized as heterogeneous and overlap each other. The oppositional young child may develop more severe conduct problems or delinquent behaviour as time goes on.

To qualify as oppositional defiant disorder, the behaviours must occur more frequently than is typically observed in children of comparable age and at a similar developmental level. Furthermore, they must lead to significant impairments in social, academic or occupational functioning.

Although oppositional defiant disorder and conduct disorder overlap in definition and presentation, they can be differentiated. Conduct disorder entails the violation of the basic rights of others, societal norms or rules (Table 2).

DSM-IV distinguishes two subtypes of conduct disorder on the basis of age at onset. This follows extensive evidence that early childhood conduct problems often persist and show a distinct risk profile. They have a markedly poorer long-term outcome than those that develop for the first time in the teens.² In childhood-onset type, at least one of the 15 behaviours (Table 2) must appear before the age of 10. In adolescent-onset type, none appears before the age of 10 years.

Prevalence

Community studies have consistently shown that the prevalence of conduct disorders is high. A recent UK study based on a national sample of 10 000 children aged 5–15 years, found that 5–10% met DSM diagnostic criteria for oppositional defiant disorder.³ The more serious problems included in the diagnosis of conduct disorder have a comparable prevalence of 2–9%.⁴ Prevalence is higher in low socioeconomic status groups.⁵

Conduct problems are the most common reason for referring young children to mental health services, accounting for 30–40% of referrals to Child and Adolescent Mental Health Service (CAMHS).^{6,7}

Oppositional defiant disorder

DSM-IV states that it is characterized by the frequent occurrence of at least four of the following, that persist for at least 6 months:

- Losing one's temper with adults
- Arguing with adults
- Actively defying or refusing to comply with the request or rules of adults
- Deliberately doing things that will annoy others
- Blaming others for their own mistakes or misbehaviour
- Being touchy or easily annoyed by others
- Being angry and resentful
- Being spiteful or vindictive

Table 1

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Conduct disorder

DSM-IV lists the following 15 behaviours categorized under four headings. At least three must be overtly present in the previous year to meet the criteria, with one present in the last 6 months:

Aggressiveness to people and animals

- Bullying
- Fighting
- Using a weapon
- Physical cruelty to people
- Physical cruelty to animals
- Stealing with confrontation of the victim
- Forced sexual activity

Property destruction

- Fire setting
- Other destruction of property

Deceptiveness or theft

- Breaking or entering
- Lying for personal gain
- Stealing without confronting the victim

Serious rule violation

- Staying out at night or being truant before the age of 13 years
- Has run away from home overnight at least twice
- Is often truant from school, beginning before the age of 13 years

Table 2

In early childhood boys are far more likely than girls to manifest disturbance of conduct. During adolescences the proportion of girls increases markedly. There are indications that gender differences in delinquency have narrowed in recent years. Symptoms of conduct disorder may differ between females and males. Some studies suggest that girls who meet the criteria for conduct disorder have a preponderance of non-aggressive or covertly aggressive symptoms. These have been most studied in adolescent girls⁸ where common presenting symptoms include those listed in Table 3.

Aetiology

A wide variety of biological, psychological and social factors are now thought to contribute to the development and maintenance of conduct disorders.⁹ These factors overlap and, when problems are longstanding, simple unidirectional models of causation will fail to capture the complex, cumulative nature of the processes involved. Many of the features of oppositional behaviour involve interactions of reciprocal compounding factors, e.g. a temperamentally difficult child may evoke negative responses from parents, which reinforce or exacerbate aggressive, non-compliant responses from the child.

Patterson's research¹⁰ described cycles of escalating coercive child – parent interactions in the homes of aggressive children. He felt harsh, abrasive and inconsistent parenting practices showed strong correlation with antisocial behaviour in children.

Twin studies suggest that shared environmental effects account for approximately 30% of the variance in childhood

Common symptoms of conduct disorders in adolescent females

- Chronic violation of rules at school
- Chronic lying
- Underachieving academically
- Substance misuse
- Non-confrontational stealing
- Running away from home
- Somatization (medically unexplained somatic complaints)
- Increased rates of arrest for non-violent crimes

Table 3

behavioural problems.¹¹ Though less heritable than other childhood disorders, conduct problems probably do have an inherited component, which is most important for the early-onset difficulties comorbid with hyperactivity.¹²

Neuropsychological deficits in children, such as low IQ, poor verbal skills and impairment in executive functioning, have been proposed as key vulnerability factors for early-onset conduct problems.² Characteristic patterns of social cognition, whereby some young people over-attribute hostile intent to others, are among individual correlates of disruptiveness and aggression, though their causal role remains less certain.

Alongside these individual factors, much research has shown that antisocial behaviour is more likely to develop in adverse family and social circumstances. Poverty, disorganized neighbourhoods, poor schools, family breakdown, parental psychopathology, harsh and ineffective parenting and inadequate supervision are all strong correlates of conduct disorder.¹³

Thus, many apparently environmental effects may reflect gene – environment interplay, whereby heritable child characteristics evoke particular responses from parents, or heritable parent characteristics influence the styles of parenting and models of behaviour the parent provides. At present, the research is only at the very earliest stages of teasing out how these complex processes operate.

Comorbid difficulties

Many pre-pubertal children diagnosed with oppositional defiant disorder and conduct disorder show predictable comorbidity^{14,15} (Table 4).

Deficits in verbal skills have long been recognized among children with conduct disorder. These can commonly manifest as specific reading difficulty or as problems in language development. As the child progresses through the educational curriculum, the demands for tasks that require verbal skills escalate. This may contribute to a breakdown of the relationship between the child and the school.¹⁶

Specific language impairment may be associated with impairments in social cognitions. These include the inability to understand humour or sarcasm, and another's mental state or intentions. Clinical observations of children with these social difficulties suggest they often use inappropriate aggressive responses to perceptions of victimization. These are particularly

Co-morbidity with conduct disorders

- ADHD
- Depression/anxiety
- Self-harm
- Substance misuse
- Post-traumatic stress disorder (PTSD)
- Learning disability
- Tourette syndrome
- Autistic spectrum disorder

Table 4

common during unstructured school breaks or poorly supervised peer interactions. This can lead to rejection by peers and teachers so children with conduct disorder face misunderstanding, punishment and alienation from school or other social environments. Parents and schools can resort to avoiding any situation which will result in a public conflict scenario. This in turn produces feelings of rejection, sadness and isolation in the child when they are not allowed or invited on ordinary social events such as school trips, peer birthday parties or even family gatherings.¹⁷

Attention-deficit hyperactivity disorder (ADHD) is a key differential diagnosis of conduct disorder and may reflect abnormalities or immaturity of prefrontal cortical functioning. It is because of this deficit that the child appears, for instance, distractible and impulsive. The 'life course persistent' conduct disorder of early-onset and sustained course may be characteristically linked to ADHD.¹⁴ There can be a failure to develop an anticipation of consequences of actions, so serious risk-taking behaviour may result if the deficit persists.

Outcomes

There is now little doubt that the long-term outlook for many children with conduct problems is poor.¹⁸ Longitudinal studies in a number of countries, and in different historical eras, have documented a litany of adverse adult outcome. These include persistent antisocial behaviours and offending, problems with employment, difficulties in interpersonal relationships, both social and intimate, high rates of alcohol problems and substance use, increased risk of mental health difficulties and compromised physical health. Though children with oppositional defiant disorder and conduct disorder frequently come from disadvantaged backgrounds, these poor outcomes are not simply a function of social adversity.

Criminology statistics reflect the long-term consequences of conduct problems. UK analyses of Home Office statistics show that in 1994 boys and young men between the ages of 10 and 20 committed 42% of all indictable offences.¹⁹ These figures probably underestimate the prevalence of youth criminal activity as a large number of offences are not reported or recorded by the police.

Adult development is compromised for boys and girls in different ways. Outcomes for boys tend to be characterized by continuing antisocial behaviour, risk of substance misuse and problems in employment, while for girls the central features may

be early pregnancy, unsupportive relationships, depression and parenting difficulties. All too often young people with conduct problems seem attracted to one another as partners in early adulthood and form volatile relationships. This way, intergenerational continuities in antisocial behaviour are likely to be high.

Conduct problems in children increase demands on multiple services, including social services, education, health and juvenile justice services. There are significant extra costs for the families with a child with a conduct disorder because of their destructiveness, and loss of employment opportunities for parents. Preliminary findings from a study of the lifetime costs found that an individual who has a conduct disorder at age 10 costs over £100 000 more in services, up to age 28 years, than one who does not.¹⁷

Assessment

Antisocial behaviour affects many aspects of a child's and young person's development. Therefore, an assessment needs to be multifaceted, involving information gathering from school and other agencies, as well as the carers and child themselves.

Of all the parties involved, the child may be the least concerned about their difficulties and may resent any attempts to help. The child's perspective is crucial, both for a complete account of the behaviour and also for an understanding of family or other problems that may exacerbate their behaviour. Drawing on a range of sources can establish the characteristics of the behavioural problems, including impulsive or risk-taking behaviours. A picture is gained of any possible environmental variation, such as the behaviours being worse at home or school, and how much other aspects of functioning are impaired.

Because co-morbid conditions can affect the course of conduct problems and may require specific interventions, it is important to enquire about symptoms of ADHD, anxiety and depression, as well as antisocial behaviour.

It is essential to ascertain an understanding of the child's interpersonal skills and approaches to conflict negotiation. The latter can be obtained by parental descriptions of examples of oppositional behaviour. A detailed 'blow by blow' account of a recent episode of difficult behaviour is a useful tool in gaining an understanding of how behaviours can arise and attempts to resolve them. Observations in the clinical setting when parent and child are engaged in a joint task, e.g. tidying up toys, can be a helpful addition to the assessment.

The quality of peer relationships, as well as the types of activities undertaken with peers, is important in formulating treatment plans. Direct observations of peer interactions, e.g. in the playground, can aid the understanding of distorted social skills.

Several checklists can be used for rating symptoms in children. One of the most commonly used is the child behaviour checklist,²⁰ which has between 100 and 113 items that describe specific behavioural and emotional problems.

Ideally diagnostic interviews should be accompanied by a full psychometric assessment. Reports from school need to cover not only the impact of the behaviour in the classroom but also academic strengths and weaknesses, and the presence or suspicions of specific learning difficulties.

Assessment of family relationships and parenting styles is crucial. Parental discord, especially around how to respond to the

child's behaviour, is important to illicit and may not be readily offered if only one parent attends. It is therefore important to ascertain if both parents and other carers, such as grandparents and child minders, have a consistent approach to the challenging behaviour of the child.

It is also essential to gain a picture of the affective tone of the family interactions. Unfortunately, by the time carers present to services they may view the child in a predominantly negative light, because of the challenging behaviours. However, it is important to look for evidence of warmth, approval and sensitivity to the child's needs. Professionals assessing a child or adolescent for a conduct disorder need to be vigilant, as they would with any child, to indicators of neglect and maltreatment.

Interventions

Conduct disturbances are difficult to treat, especially if long-standing. To be effective, any intervention needs to address the full range of the child's difficulties, at home, school and the wider community, in a developmentally appropriate way. Because conduct problems often arise in disadvantaged families, broader family problems may also need to be targeted. Engaging carers is crucial but is at times challenging. Conduct disorders often take a chronic course and rarely respond to short-term interventions.

There is now a strong argument that interventions should be informed by the increasing understanding of the psychopathology underlying conduct problems.^{21,22} Therefore, for early-onset conduct disorder or oppositional defiant disorder, interventions should focus on psychoeducation and support for parents and school, to avoid reinforcing undesirable behaviours. Problems with language, literacy and the ability to cope with peers should also be identified and addressed. Treating co-morbid psychiatric conditions such as ADHD or depression is crucial.

Parent management training

For young children (under 8 years), parent management training has the strongest support from evaluative research.^{22,23} Although there are many types of parent management training, the underlying rationale for all is based on the general view that conduct disorders are inadvertently developed and sustained in the home by maladaptive parent-child interactions. Parent management training refers to the procedures in which parents are trained to interact differently with their children. It involves the teaching of parents, individually and in groups, to pay attention to and reinforce desirable behaviours, and to practise effective strategies for dealing with unwanted responses. Role play and video feedback are used to facilitate new styles of parental responses.

Accumulating evidence shows that parent training programmes may be applied to a wide range of conduct problems and effectively delivered in various settings. NICE guidance²³ was published in 2006 on parent training/education programmes in the management of children under 12 years with conduct disorders. It identified 16 reviews that assessed the effectiveness of one or more parent training programmes. From this analysis, the recommendations listed in Table 5 were offered.

NICE guidance on parent training/education programmes in the management of children under 12 years with conduct disorder²³

1. Group-based parent training/education programmes are recommended in the management of children with conduct disorders.
2. Individual-based parent training/education programmes are recommended in the management of children with conduct disorders only in situations where there are particular difficulties in engaging with the parents or a family's needs are too complex to be met by group-based parent training/education programmes.
3. It is recommended that all parent training/education programmes, whether group or individual based, should:
 - Be structured and a curriculum informed by principles of social learning theory;
 - Include relationship-enhancing strategies;
 - Offer a sufficient number of sessions, with an optimum of 8-12, to maximize the possible benefits for participants;
 - Enable parents to identify their own parenting objectives;
 - Incorporate role play during sessions, as well as homework to be undertaken between sessions, to achieve generalization of newly rehearsed behaviours to the home situation;
 - Be delivered by appropriately trained and skilled facilitators who are supervised, have access to necessary ongoing professional development and are able to engage in a productive therapeutic alliance with parents;
 - Adhere to the programme developer's manual and employ all of the necessary materials to ensure consistent implementation of the programme.
4. Programmes should demonstrate proven effectiveness. This should be based on evidence from randomized controlled trials or other suitable rigorous evaluation methods undertaken independently.
5. Programme providers should also ensure that support is available to enable the participation of parents who might otherwise find it difficult to access these programmes.

Table 5

Failure to benefit from parent training/education programmes is associated with parental disadvantage, lack of parental perception of a need for an intervention, parental mental health problems, especially alcohol and drug problems, personality difficulties and depression. Co-morbidity in the child, such as untreated ADHD, language disorders and learning difficulties, may also reduce the efficacy of this kind of programme.

The increasing recognition of social skills deficits has generated a range of treatments that focus on the distorted approaches to social events by young children with conduct problems. Cognitive problem-solving skills training which is designed to improve the child's understanding of interpersonal situations and extend their repertoire of effective responses has also been shown to have some positive effects, if offered alongside parent training programmes.

Interventions effective for adolescents

Meta-analysis of treatment approaches to conduct disorder in adolescents has produced different findings in relation to efficacy.

Multisystemic therapy (MST) is the most promising intervention for adolescents with serious conduct disorders.²⁴ This approach recognizes the multidimensional nature of serious antisocial behaviour. It draws on a broad spectrum of techniques to address individual, parental, family and peer relationship problems. It uses multiple interventions in combinations directed by the clinical picture.

The main treatment interventions include systemic and structural family therapy, parent training, marital therapy and supportive psychotherapy related to interpersonal problems and social skills components, as well as case management where the therapist acts as an advocate to outside agencies, for the young person and family.

The main goal of MST is to give parents the skills and resources needed independently to address the difficulty of raising adolescents, while also empowering the young person to cope with familial and extra familial problems. There have been a number of trials of MST which have shown improvement of family functioning and reduction in the rate of recidivism.²⁴

A functional family therapy (FFT) approach assumes that an adolescent's problem behaviour is serving a necessary function in the family. Such functions include regulation of support and intimacy or regulation of distance between family members. The treatment focuses on the interactional aspect of the family processes as well as behavioural and cognitive dysfunctions. The goal of treatment is the achievement of a change in patterns of interactions and communications, in order to promote adaptive family functioning. Highly conflictual family interactions are a primary target of the intervention.

FFT is unusual among psychosocial therapies for children and adolescents in having a respected body of research to support its efficacy. However, effective delivery of this treatment requires considerable training and supervision. This may explain why, despite the relatively well-established manner of this treatment approach, its use is limited.²⁵

Interventions for conduct disorders that seem less effective

Social skills training and programmes to improve problem-solving skills, often referred to generically as 'anger management training', seem to have high face validity and are often pursued as a solution by parents and schools. However, they have not been demonstrated to be effective in any age group as a stand alone intervention. Trials of social skills training in adolescents with antisocial behaviour reveal there is no evidence for the skills that can be generalized beyond the treatment setting, and therefore these have limited impact on the delinquent behaviour.²⁵

Some evidence has emerged that placing aggressive or antisocial children and adolescents in group therapy may be less than useful or even exacerbate the unwanted behaviours. When youngsters with conduct disorders are placed together, in the absence of children with pro-social behaviour, i.e. without conduct disorder, peer bonding to deviant group members may occur and reinforce antisocial attitudes, values and behaviours. Ironically, the current management of children and adolescents in schools and youth offending facilities often emphasizes group interventions for conduct disorder.

Concluding remarks

Conduct disturbances in children and adolescents have significant effects on individual's mental and physical health and are an enormous strain on the resources of child health and welfare services. Advances in the understanding of the pathways leading to conduct disorder and delinquency have increased in recent years. More elaborate and effective interventions have been developed as a result. They have also given a sense of optimism to a previously 'heart sink' aspect of childhood behaviour difficulties.

If relevant interventions are available to children, parents and schools early enough, the developmental pathways for these children can be more positive. 'Early intervention' programmes offered to 'at-risk' families when the children are under 5 years may even prevent youngsters developing conduct problems and promote the healthy emotional wellbeing that accompanies the development of pro-social behaviour. The recently published NICE guidelines are an attempt to clarify what kind of parent management works. If applied in a strategically targeted approach, with adequate resources, this might not only reduce rates, but also improve the quality of life and opportunities for a significant number of young people. Further positive effects could be on rates of substance misuse, teenage pregnancies, suicide and crime. ◆

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Practice points

- Conduct disorders in children and adolescents produce enormous challenges to parents and teachers, and represent a significant burden to wider society, both in social and financial terms
- Conduct disorders need to be distinguished from the ordinary emergence of oppositional or challenging behaviour in young children as part of their psychosocial development
- Community studies have shown the prevalence of conduct disorders to be as high as 10% up to the age of 15 years. They represent the most common reason for referring young children to Child and Adolescent Mental Health Service
- The long term outcome for conduct disorder, in the absence of effective interventions, is poor
- Parent management training has the strongest evidence base, as an effective intervention for conduct disorder, in children under the age of 12 years
- In adolescents with conduct disorder, multi-systemic therapy and functional family therapy have been shown to be effective
- Social skills training or 'anger management' as stand alone interventions have poor evidence credentials, while group therapy may exacerbate the unwanted behaviours.

Stress and post-traumatic stress disorder

Guinevere Tufnell

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Abstract

There is growing awareness and concern about the high prevalence of traumatic experience and its long-term impact on the physical and mental health of children. Professionals working in the field of child health have a crucial role in identifying children at risk and in providing support for resilience and recovery. This review provides an update on current research and good practice, as well as practical advice about the available evidence-based interventions and treatments.

Keywords acute stress disorder; disaster; post-traumatic stress disorder (PTSD); stress; trauma

Introduction

Paediatricians have an important role in the early recognition of stress symptoms, which may develop into post-traumatic stress disorder (PTSD). Some of their patients will be victims of stressful events such as life-threatening illnesses, road traffic accidents, natural disasters or terrorist attacks. Others will develop symptoms as a result of the chronic trauma of witnessing domestic violence, or experiencing physical or sexual abuse. Paediatricians who are well-informed and alert to the possibility of trauma symptoms can do a great deal in a preventative capacity to ensure that appropriate supportive care is put into place. If unrecognized and untreated, trauma symptoms can persist for many months or years, impairing quality of life and development.

Definition

Following stressful events it is not unusual for survivors, including children, to experience transient symptoms such as difficulty sleeping, troubling memories and thoughts, and some disturbance in everyday functioning. An acute stress disorder¹ may be

present if the stressor was highly threatening and the reaction continues for days or weeks. When the disturbance lasts longer than 1 month and causes significant distress or impairment, it may meet criteria PTSD¹ (Table 1).

Diagnostic criteria

Symptoms of PTSD cluster into three broad categories: re-experiencing, avoidance and hyperarousal. Children over the age of 7 or 8 years display the same kinds of symptoms as adults. Scheeringa et al² have proposed alternative, more developmentally appropriate criteria for pre-school children. These criteria rely less on the child's verbal descriptions and more on what can be observed. Re-experiencing often takes the form of post-traumatic play in this age group, characteristically displaying a re-enactment of part of the trauma. Play may have a compulsive, rather stereotypic quality, and is less elaborated and imaginative than usual play. Verbally competent young children may recall fragments of the experience not necessarily showing any associated distress. There may be nightmares without an obvious trauma-specific content. Emotional numbing can take the form of constricted play patterns, social and emotional withdrawal. Regression, in the form of a loss of previously acquired skills (e.g. language, toilet training) or more immature behaviour can occur. Night terrors, sleep difficulties, general fearfulness and aggression are common.

Alan was 3 years old when he was involved in a car accident in which his father was killed. He sustained minor injuries but was alone in the dark, cold and rain for a long time before being rescued. A month later, he was easily upset, clingy, and suffered from severe night terrors. He was reluctant to travel in cars and his play showed preoccupation with themes of car crashes and destruction.

Irina, aged 12, was a refugee from Albania. She had witnessed the torching of her home and village by soldiers who committed many atrocities against friends and neighbours. She was lucky to escape alive and unharmed. She suffered from intense flashbacks and nightmares, drenching sweats, headaches and tormenting feelings of guilt and depression.

Epidemiology

Community samples suggest a lifetime prevalence of PTSD of 4–12% and a point prevalence of 1%.³ Refugees and children exposed to the trauma and losses of war will have much higher rates of PTSD, around 30–40%. It is estimated that one-third of children will develop PTSD after road traffic accidents. After a natural disaster, depending on the circumstances, very high rates (40–50%) of PTSD can be expected initially, and approximately one-third if untreated will continue to exhibit PTSD at 1 year, and as many as one-third will still have PTSD 5–8 years later.⁴

A particularly important vulnerable group for paediatricians to be aware of is children who have had traumatic medical experiences. High rates of PTSD (up to 21%) have been found in children who have been in a paediatric intensive care unit (PICU), as well as in their parents.⁵ Stoddard et al⁶ found that one-third of children will develop PTSD after severe burns. Children undergoing organ transplant are also vulnerable.⁷

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DSM diagnostic criteria for post-traumatic stress disorder

A Exposure and response

The person has been exposed to a traumatic event in which **both** of the following were present:

- The person experienced, witnessed or was confronted with an event(s) that involved actual or threatened death, or serious injury or a threat to the physical integrity of self or others
- The person's response involved intense fear, helplessness or horror. Note: In children, this may be expressed instead by disorganized or agitated behaviour

B Re-experiencing

The traumatic event is persistently re-experienced in one (or more) of the following ways:

- Recurrent and intrusive, distressing recollections of the event, including images, thoughts or perceptions. Note: In young children, repetitive play may occur, in which themes or aspects of the trauma are expressed
- Recurrent distressing dreams of the event. Note: In children, there may be frightening dreams without recognizable content
- Acting or feeling as if the traumatic event were recurring (includes a sense of reliving the experience, illusions, hallucinations and dissociative flashback episodes, including those that occur on awakening or when intoxicated). Note: In young children, trauma-specific re-enactment may occur
- Intense psychological distress at exposure to internal or external cues that symbolize or resemble an aspect of the traumatic event
- Physiological reactivity on exposure to internal or external cues that symbolize or resemble an aspect of the traumatic event

C Avoidance

Persistent avoidance of stimuli associated with the trauma and numbing of general responsiveness (not present before the trauma), as indicated by three (or more) of the following:

- Efforts to avoid thoughts, feelings or conversations associated with the trauma
- Efforts to avoid activities, places or people that arouse recollections of the trauma
- Inability to recall an important aspect of the trauma
- Markedly diminished interest or participation in significant activities
- Feeling of detachment or estrangement from others
- Restricted range of affect, e.g. unable to have loving feelings
- Sense of a foreshortened future, e.g. does not expect to have a career, marriage, children or a normal lifespan

D Hyperarousal

Persistent symptoms of increased arousal (not present before the trauma), as indicated by two or more of the following:

- Difficulty falling or staying asleep
- Irritability or outbursts of anger
- Difficulty concentrating
- Hypervigilance
- Exaggerated startle response

E Duration of the disturbance

Duration of symptoms in criteria B, C and D is more than 1 month

F Disability

The disturbance causes clinically significant distress or impairment in social, occupational or other important areas of functioning.

Table 1

PTSD does not only occur after single traumatic events. It has also been found in survivors of chronic maltreatment.^{8,9}

Although the incidence of PTSD is high, it is also worth remembering that many children will experience troubling post-traumatic symptoms that do not reach the threshold for a diagnosis, and yet have a significant negative impact on quality of life.

Pathogenesis

In situations of acute stress and danger there is an automatic psychophysiological response mediated by the autonomic nervous system, which enables us to respond (fight, flight or freeze). As part of this response, the hypothalamic-pituitary-adrenal (HPA) axis releases noradrenaline (norepinephrine) and cortisol into the bloodstream, prolonging the body's capacity to cope with stress. The whole organism switches into high alert, focusing on the business of survival; non-essential physical and psychological functions are suspended. When the event is over, normal physical and psychological functioning gradually resumes. However, the experience of trauma can have enduring effects on a child's expectations and beliefs long after the event.

The symptoms of post-traumatic stress occur when the emergency response system outlined above has been conditioned, as a result of the experience, to respond to stimuli which trigger memories of the event. However, the trauma memories that are recalled are very different from normal memory. Normal event memory is stored in the brain as information which is experienced as having occurred in the past; it includes contextual information and can be retrieved at will, in narrative form, e.g. 'I remember going to my sister's wedding last week...' Memories of this kind have been termed verbally accessible memory (VAM).¹⁰ In contrast, trauma memories are stored in the form of the original sensations, are experienced as 'it is happening again, now....' (i.e. re-living the event) and lack a narrative form. They are retrieved involuntarily, being triggered by environmental cues, and have therefore been termed situationally accessible memories (SAM).¹⁰ Normally, event information entering the amygdala and the thalamus is converted to VAMs by means of processing in the hippocampus. However, in conditions of stress, cortisol and noradrenaline (norepinephrine) inhibit the hippocampus from processing the event information in the normal way, so that storage occurs without the addition of the contextual information characteristic of VAMs.

Chronic or repeated traumatization can lead to ongoing HPA-axis dysregulation, contributing to long-term adverse effects on mental and physical health.¹³

Natural history

Not all acute stress reactions will develop into PTSD and, of those that do, 10–15% will be of delayed onset, developing 6 months or more after the event. However, quite often symptoms will resolve after a few days or weeks. The outcome depends on a number of risk and resiliency factors:

- nature of the event and the child's perception of the degree of threat
- age at exposure (greater vulnerability at a young age)
- parental mental health problems (especially PTSD and depression)
- pre-trauma psychopathology
- female gender (increases likelihood of PTSD)
- family/social/cultural support (good support increases resilience).

If untreated, PTSD symptoms can persist for years. Follow-up of survivors of the Aberfan disaster after 33 years found many to be suffering from PTSD.¹¹

Traumatic experience is often accompanied by bereavement and other significant losses which may involve major disruptions in a child's living situation. The traumatic loss of a loved one, especially if unexpected, can lead to emotional numbing or intrusive thoughts and images, which can make it difficult for normal grieving to take place.

American research with Vietnam veterans has shown that extreme and protracted stress, such as occurs in war, can have enduring and debilitating effects. Adjustment difficulties, substance abuse and mood disorders are common. If children are exposed to such conditions, or to chronic maltreatment, there are similar consequences, as well as long-term effects on many aspects of development.^{12–14}

Assessment

It is useful to begin an assessment of children in the presence of their parents. Questions about the child's current circumstances, general development and functioning can be a good place to start. The interviewer should inquire directly about all the symptoms listed in the DSM-IV criteria or in a standardized trauma questionnaire. Children will not necessarily volunteer information and may be reluctant to disclose information in front of parents for fear of distressing them. Parental accounts may not be completely reliable: they may themselves be traumatized and may underestimate the degree to which their child has been affected. Interviewing the child individually is therefore important.

Withdrawn, avoidant or dissociative behaviour can make it difficult to communicate with traumatized children, so that assessments may take a long time. Clinical observations are important: increased startle reflex or other hypervigilant responses, lapses in concentration or dissociative spells may be observed during the interview. With younger children, play or drawing may be their preferred means of expressing themselves. At the end of any interview about a traumatic experience, a child should be helped to 'wind down' by summarizing and reviewing what was said and praising them for their courage in discussing such painful events.¹⁵

Parents will often be able to provide useful background history. If they have shared in the traumatic experience they should be screened for PTSD. Their mental health and coping skills will

have a significant impact on their ability to contain their child's anxiety and provide the support that the child needs.

The use of a screening instrument, such as the child version of the Impact of Events Scale (CR-IES),¹⁶ is a useful adjunct to a clinical assessment involving single traumatic events. It is also recommended for routine use by paediatricians after a medical or other experience known to be traumatizing, such as organ transplant, PICU or a road traffic accident. Self-report questionnaires are only considered useful in children over the age of 7 years.

Differential diagnosis

Reactions to stress can take many forms, PTSD being one of them. There is a spectrum of responses, ranging from a normal stress response to a self-limited adjustment disorder, to a range of psychiatric disorders including but not limited to: anxiety, depression, dissociative states or somatization disorder.

Co-morbidity with PTSD is very common, particularly anxiety and depression, which should be looked for specifically. Trauma symptoms may also mimic other conditions. Busy clinicians who are not alert to this may be tempted to diagnose attention-deficit hyperactivity disorder (ADHD) in a child with hyperarousal and poor concentration, missing the connection with traumatic experience. Dissociative episodes can be mistaken initially for petit mal epilepsy. Children with a background of abuse and neglect can present with a wide range of psychological problems, including self-harm, substance abuse, conduct problems and aggression, often leading to the development of personality disorder as an adult. Exploring the possibility of traumatic experiences is vital, as it has important therapeutic implications.

David was 6 when he was referred for assessment of behaviour problems that were causing major problems in the classroom. He had been diagnosed as having ADHD and was taking Ritalin twice daily, but the effect of this was wearing off very rapidly. A careful psychosocial history revealed that he had also been witness to significant domestic violence between his parents. David's behaviour improved with incremental adjustments to his dose of Ritalin and support for his mother in understanding and managing the effects of past trauma.

Management

Paediatricians have a crucial role in enabling children and their parents to recover.¹⁷ Simple reassurance, information and advice are needed:

- helping to restore a sense of safety
- providing information about help available
- normalizing trauma symptoms and reassuring about recovery
- advising on the need to restore basic routine and consistency
- suggesting practical strategies for dealing with troubling symptoms such as hyperarousal and sleep disturbance
- encouraging family to identify and access sources of support within family and community
- promoting self help by providing access to useful resources such as leaflets and websites (see below).

This approach, described as 'psychological first aid', may be sufficient to enable recovery to occur. It is, however, important

that the paediatrician remains involved in a monitoring role. The child or adolescent can be asked at follow-up appointments whether they continue to be preoccupied by the traumatic event(s) or troubled by symptoms. The use of a screening questionnaire such as the CR-IES can be particularly helpful here. Scores over 17 for the avoidance and re-experiencing subscales indicate the likelihood of a PTSD diagnosis.

If symptoms are very troubling and persist for more than 1 month, referral to a specialist will need to be considered. Here again the paediatrician's role is very important. Families and young people can be reluctant to access Child and Adolescent Mental Health Services (CAMHS), and a supportive relationship with a paediatrician can greatly assist the referral and engagement process.

Prevention, treatment and prognosis

Establishing a feeling of safety and security for the child and their parents is of paramount and immediate importance. This principle is equally applicable after a natural disaster or in circumstances where a child has been witnessing severe domestic violence.

The importance of each hospital and region having an up-to-date disaster plan, which makes provision for the mental health needs of all those affected, was underlined by recent experience following the London bombings in 2005. Children may be caught up in natural disasters such as flooding or earthquakes or in man-made events such as the terrorist bombing in Omagh 1998. In some, such as the school shootings in Dunblane and in the Beslan terrorist attack, children have been specific targets. It is therefore very important that the needs of children are not overlooked by planners.

Psychological first aid (see above) carried out by a paediatrician or other professional is an important preventive measure, and may be all that is required. 'Debriefing', a technique developed in the Vietnam war, is now considered controversial and is not routinely recommended.³

Specialist psychological treatment may be required in a small number of cases. It is now established that brief trauma-focused psychological therapy is as effective in children as it is in adults, even in very young children.³ Treatment of trauma involves three phases: stabilization, desensitization and re-integration. Stabilization involves enhancing coping mechanisms, building self-esteem, improving control over emotional responses, and building or reinforcing a stable and effective support network before proceeding to trauma-focused desensitization work. In simple cases, all three phases can be completed within two to eight sessions. In complex cases, such as in developmental trauma, a great deal of 'stabilization' work is often required and treatment is longer.

Cognitive behavioural therapy (CBT) is a well-established trauma-focused treatment with a solid evidence base in children and young people. This approach involves helping the child to recall the distressing events (imaginal exposure) in such a way as to reduce distress (desensitization) and enable symptoms to be mastered (cognitive restructuring). Very young children are unlikely to benefit from formal CBT, but they can be helped with similar approaches using play, drawing and narrative techniques.

Eye movement desensitization and reprocessing (EMDR) is a relatively new technique that has shown promising results so far in research with traumatized adults. To date, there are few controlled trials in children. Symptom improvement is rapid and well-maintained, even in very young children. EMDR uses many of the same elements as CBT but relies less on homework and verbal competency, and is particularly helpful with avoidant or very young children.

Medication can be a useful adjunct as part of a multidimensional treatment approach,³ but should not be routinely prescribed. There is limited research in children. Selective serotonin re-uptake inhibitors (SSRIs) can be effective in adolescents suffering from symptoms of hyperarousal, such as poor sleep and concentration. SSRIs are also helpful in treating co-morbid anxiety and depression. Night terrors, startle responses, avoidance reactions and overactivity may respond to a beta-blocker such as propranolol or clonidine (an alpha-2 noradrenergic agonist).

George was 14 when he was mugged for the second time on his way back from school. His PTSD symptoms turned him from a popular, outgoing lad to an anxious recluse. After eight sessions of EMDR, George reported that he no longer had any PTSD symptoms and he was able to return to his previous activities. Six months later, progress had been maintained.

New developments in the field

In addition to the developments mentioned above, the following are especially worth noting.

- Concerns about the impact of trauma on the physical and mental health of children and the escalation of violence involving children in the USA has led to the setting up of the National Child Trauma Stress Network with funding provided by government. Over 50 clinics have joined and the network has provided a strong impetus for improving both services and the evidence base for effective intervention. The website provides a wealth of information and resources for both parents and professionals (see below).
- The review of DSM-IV and the field trials in relation to the diagnosis of PTSD has done much to improve the understanding of the impact of chronic interpersonal trauma/abuse on children's development across many domains, and has led to the call for a new category of developmental trauma disorder to be included in the next edition of the DSM.^{8,12}
- The brain imaging studies of Michael De Bellis and others suggest that longer duration of abuse and greater severity of PTSD symptoms are associated with smaller brain volumes.¹⁸
- The intergenerational transmission of trauma from parents to offspring has been studied by Rachel Yehuda and her team in New York. Dysregulation of the HPA axis has been found to play an important part in this.¹⁹ ◆

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FURTHER READING AND USEFUL RESOURCES

Vasterling JJ, Brewin CR, eds. The neuropsychology of PTSD: biological, cognitive and clinical perspectives. New York: Guilford Press, 2005.

www.nctsnet.org The National Child Trauma Stress Network is a major resource for parents and professionals.

www.rcpsych.ac.uk/mentalhealthinformation/youngpeople.aspx The Royal College of Psychiatrists has a useful series of downloadable factsheets for parents and young people.

Practice points

- Screening for trauma symptoms should be routine after traumatic medical experiences, stressful events and where there are concerns about possible abuse and domestic violence
- A detailed psychosocial and trauma history should be taken in conditions presenting with hyperarousal and affect dysregulation, e.g. ADHD
- Early interventions to support resilience and enhance coping can be highly effective in preventing long-term morbidity
- Referral to specialist services should be made when PTSD symptoms persist for longer than 1 month

Chronic fatigue syndrome/myalgic encephalopathy in children

A T Anbu

A G Cleary

Abstract

Chronic fatigue syndrome/myalgic encephalopathy (CFS/ME) in children is a common condition associated with considerable controversy with reference to terminology, case definition, aetiology, treatment and prognosis. This frequently results in patients and families receiving mixed messages, and significant delay in establishing a diagnosis and receiving an appropriate management programme. We review current understanding and draw attention to the best practice approach to assessment, diagnosis and management of CFS/ME in children. This, we hope, will improve planning, organization and delivery of a patient-centred, optimized and clinically-effective multidisciplinary management strategy towards children with CFS/ME.

Keywords chronic fatigue syndrome/myalgic encephalomyelitis

Introduction

Chronic fatigue syndrome/myalgic encephalopathy (CFS/ME) in children is a condition associated with considerable controversy. Issues considered fundamental to medical management of any other paediatric chronic disease, such as terminology, definition, cause, treatment and prognosis, continue to provoke debate (often very emotive) and confusion among professional and lay groups alike. This may (and frequently does) result in patients and families receiving mixed messages and having to endure often significant delay to establishing a diagnosis and an appropriate management programme. This in turn may result in mistrust, distress and even hostility towards health and other professionals, such as teachers, who are perceived as unsympathetic or unaware of the impact a child's symptoms are having on both the individual and the family. An equally significant result from such factors is the inevitable impairment of research programmes to help understand factors such as aetiology and the development of an evidence-based literature on CFS/ME.

There is an incomplete distribution of specialist and local services in the UK for children with CFS/ME, which adds complexity to the medical management of a condition that is frequently profoundly disabling, e.g. in terms of a child's ability

to function in school. The skill mix and background of health professional input to children with CFS/ME may vary widely. It is likely that many children with CFS/ME are managed in primary and secondary care, with support from tertiary units for more severe cases. It is likely many cases are unrecognized and therefore untreated. Tertiary units may find themselves unable to meet the referral demand placed upon them and, if there are limited local services to share management with, will find it almost impossible to coordinate a successful programme from a distance.

We believe CFS is a biological illness, manifest with complex interlinking between adverse thoughts, moods, emotions and physical symptoms. This review explores best understanding and management of CFS/ME in children.

Terminology

CFS/ME is the widely but not universally accepted umbrella terminology for a clinically heterogeneous syndrome characterized by persistent and disabling fatigue. This is the term adopted by the Royal College of Paediatrics and Child Health (RCPCH) and is used in our unit.¹ There are some who consider CFS and ME as distinct entities. Others may classify CFS/ME under the terms 'medically unexplained symptoms' or 'functional somatic disorders'. Historically, post-viral fatigue syndrome has also been used. The choice of terminology may depend upon the training and speciality background of the health professional.

Key message: We would support the adoption of a universally accepted terminology in order to reduce confusion among patients and professionals and to facilitate better case ascertainment for the purpose of studies.

Definition

There are several published definitions of CFS/ME in adults. Diagnostic criteria from the Communicable Disease Center, USA utilize a time period of persistent symptoms of 6 months duration.² Other definitions are summarized in the RCPCH guidelines.¹ There are currently no diagnostic criteria for CFS/ME in children. The RCPCH guidelines utilize a definition that removes the requirement for symptoms to have persisted for a defined time period, recognizing the importance of establishing the diagnosis as quickly as possible. This definition for CFS/ME is shown in Table 1. The other 'classical' and frequently reported symptoms referred to in the RCPCH definition are shown in Table 2.

Definition of CFS/ME in children in RCPCH guidelines

Generalized fatigue (i.e. causing significant disruption of daily life) persisting after routine tests and investigations have failed to identify an obvious underlying 'cause'. In CFS/ME the fatigue is likely to be associated with other 'classical' symptoms, such as difficulty in concentrating and disturbed sleep patterns, and is classically exacerbated by effort (both mental and physical).

Table 1

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'Classical' and frequently reported symptoms in CFS/ME other than fatigue

Other 'classical' symptoms	Other frequently reported symptoms
Severe malaise	Feeling too hot or too cold
Headaches	Dizziness
Memory and concentration impairment	Cough
Sleep disorder	Eye pain/photophobia
Depressed mood	Visual or hearing disturbances (hyperacusis)
Myalgia at rest and on exercise	Weight loss or gain
Nausea (often pervasive) and anorexia	Muscle weakness
Sore throat	Lack of energy for usual activities
Tender lymph nodes	Diarrhoea
Abdominal pain	
Arthralgia	

Table 2

Establishing the diagnosis

The differential diagnosis is broad and can be approached with a 'surgical sieve' methodology. Making a correct diagnosis requires consideration and exclusion of a broad range of potential disorders (Table 3). As with most paediatric chronic disease, prompt diagnosis will be possible in the majority by careful history, physical examination and rapid access to guided investigations.

Key message: As a number of the diagnostic possibilities are associated with significant morbidity and some may be potentially life-threatening, the diagnosis of CFS/ME should be established by a paediatrician with the relevant training and expertise necessary to exclude such disorders. If there is diagnostic doubt, relevant paediatric sub-specialists will need to be consulted, and who should always be made aware that CFS/ME is a possible diagnosis.

Multidisciplinary assessment

In our service the initial assessment is made by a team comprising a paediatrician, clinical nurse specialist, physiotherapist and either a child psychiatrist or psychologist. This can be very daunting for the young person and family, and in many cases a full understanding of the impact of the disease can only be made over several subsequent visits. Many young people will experience an increase in symptoms after visiting the hospital and will need to be reassured that this can be managed. Generic health issues such as recreational drug misuse, alcohol intake and sexual activity should be considered where appropriate, and we encourage teenagers to engage with professionals independently of parents for part of many sessions.

Psychological assessment is considered by some stakeholders to be controversial. We believe it is essential, although in many

Potential 'checklist' for differential diagnoses associated with fatigue

Inflammatory	Juvenile idiopathic arthritis Connective tissue disorders Inflammatory bowel disease
Endocrine	Diabetes mellitus Hypothyroidism Hypoadrenalism
Primary psychiatric disorder	Organic depressive illness Childhood schizophrenia Anxiety disorder
Infection	Chronic infection – potential association with immunodeficiency Viral ('post-viral syndrome')
Neurological and neuromuscular disorders	Demyelinating disorders Myasthenia gravis Muscular dystrophies
Malignancy	Haematological Neuroblastoma CNS tumour
Haematological disorder	Iron-deficiency anaemia (may co-exist with CFS/ME)
Fabricated or induced illness	

Table 3

cases this is done by a clinical nurse specialist or physiotherapist with support from either a child psychiatrist or psychologist. Clues suggesting psychiatric illnesses like major depression or anxiety should be actively sought. Sufficient opportunities to explore other associated psychological co-morbidities, including school phobia, social withdrawal, personality problems and emotional liability, should be given, and each may need specific management.

As a minimum we perform a baseline screen of investigations (Table 4), but this must always be modified according to the nature and extent of the clinical features.

Explaining the diagnosis

As there are no pathognomic investigations or biomarkers, explaining the diagnosis can be a very challenging process. We sense many young people and families are frustrated and at times angry that such a disabling condition cannot be defined in greater scientific detail. Many young people already report feelings of isolation from their peers who 'do not understand' how they feel, and we frequently hear reported comments such as 'you look ok, so why are you not able to come to school?'

To explain the symptoms of CFS/ME, we use a framework outlining failure of the normal neuroendocrine control mechanisms, leading to a vicious cycle of events within the body. We take great care to emphasize this is not strictly evidence based, but we find it gives families a useful concept to both understand and manage the illness. This is explained in schematic form in Figure 1.

Minimum screen of investigations for CFS/ME

Urine

- Protein, blood and glucose

Blood

- Full blood count
- Renal function test
- Liver function test
- Thyroid function test
- Erythrocyte sedimentation rate (or plasma viscosity)
- C-reactive protein
- Blood glucose
- Coeliac screening
- Bone profile
- Creatine kinase
- Serum ferritin

Table 4

Epidemiology

Prevalence data for CFS/ME are heterogeneous and confusing as a result of different study methodology, case ascertainment and definition. As such, published data are difficult to interpret. Using the 1994 Communicable Disease Center (CDC) definition of CFS and self-reported symptoms, a prevalence of 0.3% is reported in a random US cohort between the ages of 12 and 17 years.³ In the UK, a retrospective cross-sectional survey reported a prevalence of 0.062% of medically unexplained severe fatigue over

3 months in 5–19 year olds.⁴ This contrasts with a prevalence rate of 0.19% in 5–15 year olds using data from the British Child and Adolescent Mental Health Survey 1999 and utilizing the CDC criteria.⁵ Long-term prospective community-based studies are needed truly to understand the prevalence of CFS/ME.

Aetiology

No single cause of CFS/ME is known, and it seems likely that the disorder is multifactorial. Evidence for a genetic component has been reviewed.⁶ Selected cases are associated with infection (Epstein–Barr virus, Q fever and viral meningitis), with no definable immunological disorder and some evidence of low levels of cortisol probably secondary to physical inactivity and sleep disturbance.⁷ In a cohort study of 16 567 babies born in 1970, higher risk of CFS/ME in adult life was associated with having a disabling condition in childhood, female sex and high social class. Higher levels of exercise in childhood were associated with lower risk of CFS/ME with no evidence that maternal psychological disorder, psychological problems in childhood, birth weight, birth order, atopy, obesity, school absence, academic ability and parental illness increased the risk.⁸ In a school population-based survey, being either highly active or highly sedentary or having evidence of low mood independently increased the risk of persistent fatigue symptoms.⁹ Clearly, for interventions to be appropriate and effective, better understanding of aetiologies is needed.

Impact of the illness

In terms of both physical and cognitive disability and also duration of symptoms, the impact of CFS/ME can be profound. In a survey in the 1990s, 42% of all medically certified long-term sickness absence from school was ascribed to CFS/ME.¹⁰ Social isolation may be equally extreme, even if using electronic methods of communication. Children often feel trapped in a vicious circle of isolation, often during their adolescent years or at the advent of secondary school.

CFS/ME in a child may in turn adversely affect parental employment and family finances. Benefits such as the disability living allowance (DLA) should be sought. We find many patients have already accessed support groups such as the Association for Young People with ME (www.ayme.org.uk), where there are many resources regarding management of symptoms.

Management

Management of CFS/ME is complex and in the majority of cases a coordinated multidisciplinary approach using the tools of pharmacological, psychological and behavioural (a biopsychosocial model) techniques is needed. Establishing a lead health professional to coordinate the management programme and actively to engage with the patient and their family is fundamental.¹¹ We designate either a physiotherapist or clinical nurse specialist from our CFS/ME team as key worker for each patient, supported by either a psychologist or psychiatrist. Patients should always be encouraged to take an active role in tailoring an *individual management plan*, and information shared with them via appropriate methods. In practice this is mostly face to face, but in our

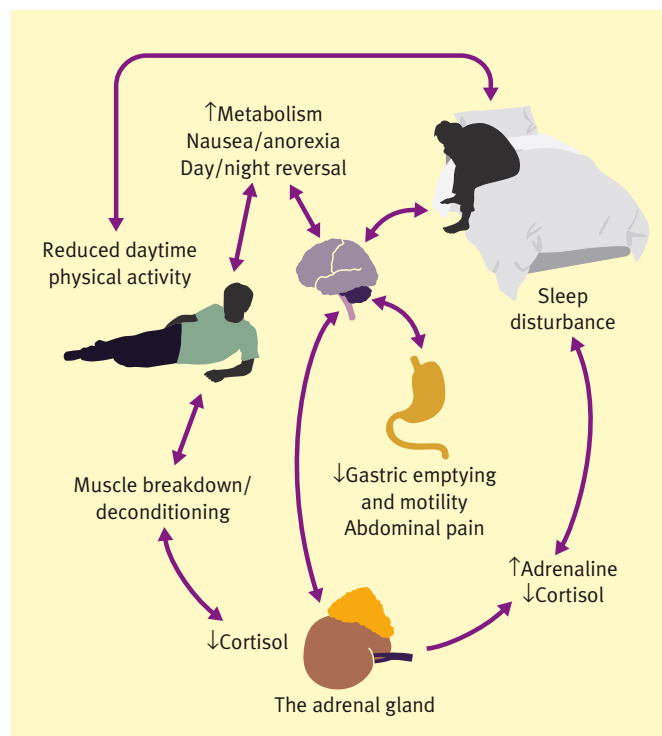


Figure 1 Schematic representation of complex interplay between central and peripheral neuroendocrine circuits.

experience methods including email and text messaging are frequently and increasingly used.

Evidence-based guidelines for the management of CFS/ME have been developed collaboratively by the RCPCH and other relevant agencies, including patients.¹ There is a National Service Framework (NSF) exemplar patient journey for CFS/ME.¹² Any professional working with young people with CFS/ME should be familiar with these documents.

Minimum standards for a management plan should include:

- Activity management that establishes a baseline level of mental and physical activity that does not exacerbate symptoms (i.e. avoid tendency to 'boom and bust'). Equally, too much rest will result in accelerated muscle deconditioning and delay recovery;
- Establish the fundamental principle of a goal and rehabilitation based approach to treatment¹³;
- Management of specific symptoms (as outlined below);
- Establish a programme of review.

Cognitive behavioural therapy

Cognitive behavioural therapy (CBT) is a technique that identifies and monitors thoughts, assumptions, beliefs and behaviors which are dysfunctional, inaccurate or simply unhelpful. Over time a behavioural element is introduced gradually and consistently to introduce a change in behaviour, e.g. an increase in activity or improvement in sleep quality. In a randomized controlled trial, CBT has shown positive and sustained improvement in physical functioning and school attendance in adolescents.¹⁴

Energy management and rest

It seems intuitive that a primary step is to establish baseline activity (physical and cognitive activities) which the child is able to undertake on a bad day without experiencing symptoms.¹⁵ This could be achieved with the use of an activity diary. It must be highlighted that *activity* includes not just physical exercise but mental activity such as reading, watching television and social contact (direct or electronic) with friends. Once the baseline is identified, the objective is for the child to perform these activities consistently for a few days or even weeks before they could be gradually increased. Regular rest periods including relaxation exercises should be included in the activity management. Once the child is able to perform consistent (week-by-week) levels of activity without experiencing symptoms, depending upon the age of the child and severity of the illness, the activities could then be gradually increased and rest periods reduced.

Graded exercise therapy

Graded exercise therapy (GET) is a physical therapy-based programme aiming to increase fitness and stamina and reduce the impact of physical deconditioning. When used as part of a rehabilitation programme with general support, GET was shown significantly to increase Wellness score and school attendance.¹³ In our experience we focus on GET as a specific goal-driven tool only after effective management of the young person's education has occurred.

Complementary therapies

Many families will ask for advice regarding complementary or homeopathic remedies. This should not necessarily be discouraged. There is no evidence base to help inform such decisions,

and we suggest patients only explore one therapy at a time, e.g. osteopathy, massage, acupuncture.¹ We always check specific homeopathic remedies for interactions with other pharmacological agents.

Management of specific symptoms

Sleep disturbance

Children frequently suffer from sleep disturbances in the form of day/night reversal, interrupted sleep, insomnia or hypersomnia. A better understanding of the sleep pattern of the child and the whole family is paramount in excluding primary sleep disorders and providing assistance and support for managing sleep-related problems. Advice should be provided on establishing good sleep routine and hygiene. This should include:

- Establishing regular sleep-wake times – waking times can be brought forward in increments (15–30 min) according to the plan;
- Avoiding prolonged sleeping and daytime sleeping, and undertaking relaxation exercises ('wind down routine' during the evening). We always insist a young person gets up and dresses out of pyjamas during the day;
- Always sleep at night in own bedroom, and avoid use of TV/computer in own bedroom if possible.

If daytime sleep is unavoidable in the early stages, allow short naps for a maximum of 40 min and no sleep after 3 pm.¹⁵ Sleep problems can also be addressed by CBT.¹

We frequently use melatonin, alone or in combination with low-dose amitriptyline (Table 5) if pain symptoms are not controlled by simple analgesics. This will help re-establish sleeping routines, but we recognize there is a limited evidence base for this strategy in children.¹

Gastrointestinal symptoms

Persistent nausea and abdominal pain is frequent and may result in dietary modification and impairment. However, adequate nutrition is essential for recovery. Bone health assessment may be particularly relevant for teenagers who undertake limited physical activity and are exposed to limited sunlight. Advice should be given on healthy balanced eating with frequent small feeds, and adequate fluid intake. Nutritional assessment and growth should be consistent with any paediatric chronic disease. Antispasmodics and antiemetics may be considered. In major

Pharmacological approach to promote sleep quality

Drug	Dose and regimen
Melatonin	<ul style="list-style-type: none"> ● Start 2 mg/dose 30 min before bed ● Increase up to 6 mg/dose ● Consider adding slow-release capsules (3 mg/dose) if waking during the night
Amitriptyline	<ul style="list-style-type: none"> ● 10 mg/dose 30 min before bed ● Helpful if additional control of chronic pain symptoms necessary

Table 5

nutritional problems, a dietician should be involved in providing support and assistance to the child and the family.¹ Rarely, admission to hospital for nasogastric feeding may be indicated if nutritional impairment becomes severe.

Pain

Pharmacological therapy with simple analgesics such as paracetamol and/or ibuprofen should be tried, although many patients will find this inadequate. Low-dose amitriptyline may be a useful adjunct at a dose of 10 mg/day, increased to 1 mg/kg/day (maximum dose 50 mg) taken 30 min before bed. Relaxation and cognitive behavioural techniques may be considered in parallel to improve pain control.¹

Education

CFS/ME can affect children at any stage of their primary or secondary school career. CFS/ME has been described in 16% of children younger than 12 years in a paediatric cohort.¹⁶ In many cases, successful return to health can only be achieved with liaison between families, health professionals and education services. We recommend home tuition for children unable consistently to attend school, and agree with others that re-integration should only be considered when the patient can manage an hour of school work daily at home plus an hour of other activity.¹⁵ Re-integration is often successful if it begins with subjects the patient enjoys or is better at, and will often begin with the lesson before lunch, gradually extending into time at school during lunchtime for socialization.

Children with CFS/ME are entitled to exam concessions, and we recommend this be requested by health professionals for their patients. A proforma for such a request is shown in Table 6.

Follow-up

Once the patient-centred, goal-oriented rehabilitative management strategy is established and commenced, frequent review will help to avoid non-compliance and deterioration in symptoms. The timing of review will vary for each child, depending upon their age, their understanding of the illness, family dynamics and the severity of the symptoms. A systematic review of symptoms, including any new ones, should be undertaken at each visit to identify early signs of relapse or setbacks. If this happens, all steps should be taken to review and modify the management plan. This may mean a step-down approach to re-establish symptom control, before gradually increasing activity again.

Prognosis

A systematic review in adult populations has reported variable outcome ranging from 5% median full recovery rate to 40% median improvement.¹⁷ However, in children the outcome is more favourable, with return to health or significant overall improvement in 80% of a UK cohort attending a GP specialist clinic.¹⁸ In such a US primary care cohort, 37% recovered completely and 43% felt well without having fully recovered.¹⁹ In a systematic review of four predominantly hospital-based studies, 54–94% of children recovered during the periods of follow-up.²⁰

Sample letter requesting examination concessions for a young person with CFS/ME

X has chronic fatigue syndrome, and is regularly reviewed by his medical team at ...

As X has this diagnosis, I would like to request exam concessions on his/her behalf. Conventional medical advice relating to young people with chronic fatigue syndrome is that they receive up to 50% extra time and are allowed rest breaks as required throughout the exam. We recommend X sits his/her exams in a room on his/her own, which would enable him/her to get up and move around without disturbing anyone else.

We also recommend that X only sits one exam per day. Exam pressure and stress will drain his/her energy levels quickly and his/her cognitive function will suffer as a result. By allowing one exam per day he/she will have a chance to rest and recharge his/her energy levels for the next day.

As chronic fatigue syndrome is a relapsing and remitting illness, I am unable to predict how X's medical condition will be on the day of his/her exams. We recommend provisions to sit exams at home be made as a precaution.

All 9 am exams will have a considerable impact on X's performance as his/her symptoms are increased in the morning and ease somewhat during the afternoon. This makes working to exam standard, particularly on consecutive exam days, extremely difficult and would have a major impact on his/her projected grades. Therefore, we are applying to have all exams scheduled for the morning changed to a later time in the day, preferable PM.

Table 6

Transfer to adult services

Despite the favourable outcome in many children and young people, there are some with persistent symptoms who will have to be managed by adult teams. Many adult services are not resourced to give patients the time with health professionals they have experienced in paediatric services, and this can be daunting for patients and professionals alike. Hence, there should be a smooth and supportive pathway of transition of care for these children and adolescents into adult services. Fundamentally, this requires the establishment of effective transition programmes, with adolescents empowered to become active partners in their health management plans, transition programme and ultimately transfer process.²¹

Conclusion

As summary for this review we have permission to publish this extract from one of our patient's, who has now recovered and is soon to embark on medical training at university.

'CFS/ME is a physically and mentally draining illness – it is erratic and unpredictable and for all these reasons makes it so difficult to deal with, treatment and management wise. You are always trying to avoid boom and bust cycles – the 'saw tooth' graph – trying to maintain set percentage ability. Education

takes a hit when a young person suffers from CFS/ME, and to maintain some supply of education, hospital and home tuition services must tailor lessons to an individual's specific needs, and if this means missing lessons, so be it, as the main priority is a person's health. But overall, greater recognition and awareness must be brought to CFS/ME, so that young people can be diagnosed and receive the correct support sooner.' ♦

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Practice points

- CFS/ME is a clinically heterogeneous syndrome characterized by persistent and disabling fatigue in children. It is usually associated with a constellation of other symptoms
- Currently there are no case definitions or diagnostic criteria specifically for children
- The RCPCH supports and promotes the fact that, 'It is a condition characterized by generalized disabling fatigue persisting after routine tests and investigations have failed to identify an obvious underlying cause'
- As there are no pathognomonic investigations or biomarkers, establishing a diagnosis requires consideration and exclusion of a broad range of potential disorders. It is recommended that this be undertaken by a paediatrician with appropriate training and expertise
- Data on the prevalence of this illness are inconsistent due to a lack of consistent case definition
- Despite significant research the exact aetiology still remains unclear
- It has significant impact on the child and in turn on the family in terms of parental employment and finances
- Management of CFS/ME is complex and usually involves an outpatient-based, rehabilitative and multidisciplinary team approach
- Symptom management should not be delayed until diagnosis is established
- CBT has proven to be an effective treatment modality
- Although the prognosis is generally favourable in children, a number of children will have to be transferred to adult services

Radiology in non-accidental head injury

Neil Stoodley

Abstract

Non-accidental head injury (NAHI) is not uncommon and has potentially serious short and long term consequences for both child and family. This review gives a brief outline of epidemiology and pathology with an explanation for the rationale behind recently published imaging guidelines. It also highlights some important potential pitfalls in interpretation.

Keywords child abuse; head injury; imaging; non-accidental head injury; radiology; subdural

Definition

Non-accidental head injury (NAHI) is head injury which occurs as a result of an abusive act by a parent or carer perpetrated on a child. Whilst the causative event is likely to be recognized as obviously inappropriate by an independent witness, it may or may not be intentional.

Epidemiology

The incidence of NAHI seems to be fairly consistent across published studies. Most such injuries occur in children below the age of 2 years, with studies suggesting annual population incidence with age in the order of up to 36 per 100 000 in children below 6 months of age, up to 24 per 100 000 in children below the age of 1 year and up to 3.8 per 100 000 in those aged 1–2 years.^{1,2} The overall incidence of NAHI is therefore similar to that of childhood cancers in an equivalent age group and is every bit as serious, with outcome studies in NAHI showing an overall mortality of around 20%, severe disability in 30–35% and moderate–mild disability in 25%.^{3–5}

Mechanism

Although the subject of some debate, the most important mechanisms involved in the majority of cases of NAHI are likely to be shaking, impact against an external surface or a combination of the two. Given that most accidental head injury involves impact head trauma, it is interesting to note the differences between accidental and NAHI in terms of neuroimaging appearances and clinical outcomes. This is likely to be due to differences in the

primary mechanisms involved and consequent differences in brain pathology.

Pathology

The two main pathologies seen on imaging in NAHI are intracranial haemorrhages and parenchymal brain injury.

Intracranial haemorrhage

The commonest type of haemorrhage seen in NAHI is subdural haematoma (SDH). Whilst SDH can undoubtedly be caused by impact head injury, in the context of impact head injury occurring as a result of typical domestic trauma, if SDH occurs it tends to be seen at a single site related to the site of impact (or occasionally diagonally opposite). In contrast, the imaging findings in most cases of NAHI are of shallow SDHs at several different sites, usually in separate intracranial compartments.^{6–9} The SDHs are unlikely to be responsible for the symptoms and signs seen in NAHI, as SDHs can be clinically silent whether they occur following birth,^{10,11} accidental trauma or NAHI,¹² and also the pattern and volume of SDHs in NAHI are similar whether the child shows few symptoms and signs or major symptoms and signs. The difference is the degree of associated brain injury, which in NAHI is usually hypoxic–ischaemic.^{13–15}

Parenchymal injuries

Focal brain injuries, such as contusions, traumatic clefts and diffuse axonal injury, are more commonly seen as a result of impact head injury (accidental or NAHI); they are unusual in injuries involving shaking which do not involve impact with an external surface.

By contrast, hypoxic–ischaemic brain injury (particularly when generalized) is an unusual finding following witnessed impact head trauma. Varying degrees of hypoxic–ischaemic brain injury are very commonly seen in NAHI, and the degree of associated brain injury correlates well with the degree of symptoms and signs seen both on presentation and in terms of short- and long-term outcomes.

Imaging investigation

Studies have consistently shown a wide variation across the UK in the investigation pathway of possible non-accidental injury.¹⁶ In an attempt to move toward a more standardized approach, the Royal Colleges of Paediatrics and Child Health (RCPCH) and of Radiologists (RCR) have recently published a joint document⁵ setting out imaging guidelines, which are, where possible, evidence based or, where evidence is (for obvious reasons) lacking, based upon widely held and reasonable clinical opinion. An imaging algorithm for suspected NAHI was incorporated into the joint RCPCH/RCR document.⁵

Imaging modalities

Skull X-rays

These should be performed in all cases. Some units omit skull X-rays if a computed tomography (CT) head scan has been (or will be) performed. This is bad practice as even quite large skull fractures may not be evident on head CT if fractures run in the plane of the scan. Frontal and lateral views should be obtained as a minimum and a Townes view may be required if there is any suggestion of occipital injury.

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Ultrasound

Cerebral ultrasound (US) scans have no part to play in the routine investigation of possible NAHI. The SDHs commonly seen in NAHI are shallow and most often occur in the posterior interhemispheric fissures, posterior fossa and over the cerebral convexities. These areas are not well seen on US.¹¹

Computed tomography

Head CT scans are the usual initial investigation and should be performed as soon as the child has been resuscitated and is stable. CT is widely available and is straightforward to perform on sick infants. CT scans are now quite fast, especially given the more widespread availability of multi-slice scanners. CT is good at demonstrating acute blood and is perfectly adequate for demonstrating the small minority of cases where intracranial pathology requires urgent neurosurgical intervention.

CT is less good at demonstrating parenchymal brain injury (both focal and generalized), although such abnormalities are often clearly demonstrated on CT in more severe injuries. CT obviously involves exposing the child to ionizing radiation with its inherent associated risks.¹⁷ However, this risk needs to be weighed against the mortality and morbidity figures for NAHI. Whilst a child who is not frankly encephalopathic is unlikely to die from an acute injury, the recognition of the true nature of their illness may well prevent them from sustaining a further and possibly more serious injury.

Magnetic resonance imaging

Magnetic resonance imaging (MRI) is an important part of the imaging pathway. Although more difficult to perform (and often interpret) than CT, it is more sensitive for the detection (and elucidation of the nature of) parenchymal brain injuries and also for showing SDH in areas not well seen on CT, such as the temporal regions and the posterior fossa. Advanced techniques, such as diffusion-weighted imaging, show evidence of hypoxic-ischaemic brain injury in most cases (even in cases where there does not appear to be any such change on standard CT and MR scans).^{18,19} In addition, it is becoming clear that spinal SDH is not infrequently seen in NAHI and, perhaps counter intuitively, early experience suggests that it may well be more common in the lumbar region than the cervical region. Although there is insufficient evidence currently to suggest the routine use of spinal MRI scans in such cases, many units that see a large number of cases are scanning the whole spine as part of the NAHI protocol.

Is it or isn't it?

These cases are all difficult and emotive, especially for those doctors who must have a direct professional relationship with parents and carers, who may, if NAHI is considered likely, be the subject of legal proceedings as possible perpetrators. Making the wrong decision either way can obviously have serious consequences. The results of imaging can only be part of the jigsaw, but it must be appropriate to investigate all potential cases in a systematic way to gather as much relevant information as possible, allowing the multidisciplinary team to come to the most reasoned decision as to the likelihood or otherwise of NAHI having occurred.

If such cases come to Court, the Court will require input from doctors in terms of explanation and interpretation of the medical evidence. Whilst the treating clinicians may well be very experienced in terms of child protection, ideally, expert review of the case should be performed by experienced clinicians not directly involved in the case. This introduces an element of peer review and allows the outside experts to approach the case afresh and with the advantage of having all the relevant material available for review retrospectively (a considerable advantage). The other main advantage is having the time adequately to consider the case rather than being involved in the constantly moving clinical process. It should also give parents and carers (and indeed involved clinicians) some confidence that those who have come to a clinical diagnosis of NAHI are not the only voices that will be heard in Court.

Avoiding pitfalls

It is important never to jump to conclusions and to discuss the case with radiologists, if necessary seeking opinions from radiologists in other centres, a process which is increasingly facilitated by the spread of picture archiving and communication systems (PACS).

Fractures

Fractures are evidence of impact injury against a hard or unyielding surface. It is not possible to assess the age of skull fractures (if there is associated soft tissue swelling, then it is probably a fairly recent injury). Skull fractures can take weeks (and sometimes months to heal). We do not know the true incidence of skull fracture following domestic trauma, as not all children have skull X-rays or scans, but we do know that fractures do occasionally occur after seemingly minor domestic falls and that there may not be any significant associated symptoms or signs following such an injury.

Most skull fractures are simple, linear parietal fractures (whether caused by accidental or non-accidental impact). More complex fractures (diastatic, depressed, stellate, involving a bone other than the parietal bone) are more commonly seen in the context of NAHI,²⁰ but this does not mean that they are all due to NAHI. The complexity of the fracture probably relates to the degree of force that caused the fracture rather than the mechanism.

Subdural haemorrhage

Short distance falls, such as toddlers falling after having pulled themselves up on furniture or pre-mobile infants (or toddlers) falling over from a sitting position, hitting their heads on the floor, can give rise to SDHs. Again, we do not know the true incidence of SDH in such events as not all infants are scanned following them. If this is the suggested mechanism of causation however, a pattern of SDH consistent with impact head injury would be expected: single-site SDH rather than multifocal haemorrhages.

Very rarely, relatively minor head trauma can lead to more serious intracranial injury. This usually occurs in the context of associated damage to arterial vessels, leading to the development of an extradural haematoma or, less commonly, a large single-site space-occupying SDH following damage to veins. Again, the pattern of haemorrhage is very different from that seen in NAHI.

Estimating the age of intracranial blood

The appearance of intracranial blood on CT and MRI scans is variable. One of the factors upon which its appearance depends is the age of the blood. On CT scans acute (recent) haemorrhage is brighter than the underlying brain and, if it persists, it becomes progressively darker: initially it has a similar attenuation to the underlying brain (subacute) and is then darker than the brain (chronic). The appearance of blood depends on other factors as well, including the volume of the haematoma and whether the patient is anaemic, but critically the assessment of the age of the blood depends on the haematoma being a discrete collection of blood and not a collection of blood mixed with other fluid [such as cerebral spinal fluid (CSF)].

It is now well recognized that dark subdural collections can be seen following acute injuries.²¹ It is believed that these occur following damage to the arachnoid membrane, which allows CSF to leak into the subdural space and either collect there or dilute any acute subdural blood present.²² Therefore, the presence of dark subdural fluid and acute subdural blood on the same scan does not necessarily mean that there is blood of two different ages or, by implication, that there have been at least two episodes of bleeding. The same is true of assessing the age of blood in terms of MR appearances: if the collection is a discrete collection of blood, then it may be possible to give some assessment of its age²³ (but even then the time window will at best be days' wide); if the acute blood is mixed with CSF or chronic subdural fluid, then it is unlikely to be possible to make a meaningful assessment.

Correlation with the clinical history

As there are no pathognomonic features of NAHI, scan appearances cannot be accurately interpreted in isolation and require correlation with the clinical history or, sometimes, with the lack of any apparent clinical history. That requires close cooperation between paediatricians and radiologists and other members of the multidisciplinary team. ♦

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Self-assessment

Questions

Case 1

A 3-year-old boy presents to A&E with fever of 39°C, HR of 140/min, capillary refill time of 3–4 s and slightly drowsy. On further questioning, it is revealed that he had returned 2 weeks previously from an East African safari, and had taken the recommended malaria prophylaxis.

- What are the two most likely diagnoses? Choose TWO answers from the following options:
 - Plasmodium falciparum* malaria
 - Plasmodium vivax* malaria
 - Bacterial sepsis
 - Influenza
 - Dengue fever
- Which of the following investigations should have priority? Chose ONE answer from the following options:
 - Malarial blood film
 - Blood glucose
 - Arterial blood gas
 - FBC
- What should be the immediate management of this child? Chose ONE answer from the following options:
 - Intravenous infusion of quinine sulphate
 - Exchange transfusion
 - Fluid restriction to 80% maintenance and intravenous quinine
 - Bolus of 20 ml/kg colloid or 0.9% saline.
- With what definitive antimalarial treatment should this child be treated? Chose ONE answer from the following options:
 - Parenteral quinine for 7 days
 - 3-day course of chloroquine
 - Parenteral artesunate
 - Intravenous quinine followed by a full course of oral medication with either mefloquine, proguanil with atovaquone, or artemeter with lumefantrine

Case 2

A 65-year-old woman visiting her family from India is admitted with a febrile illness and cough. She has a caseating lesion in the left upper lobe on chest X-ray and a sputum smear is positive for acid-fast bacilli. She had been staying with her daughter and grandchildren, who are 3 weeks, 18 months and 4 years old, respectively.

- The 3-week-old baby is well and thriving. Which management plan should be followed? Chose ONE answer from the following options:
 - Give BCG and keep under regular outpatient follow-up
 - Perform a Mantoux test and if positive, treat with full antituberculous treatment with isoniazid, rifampicin and pyrazinamide
 - Start on isoniazid 5 mg/kg and then perform a Mantoux test after 3 months' treatment
 - See in regular outpatients until he is 6 months old and then perform a Mantoux test
- The 18 month old, who had a BCG when she was 1 week old, is well when first seen. Which management plan should be followed for her? Chose ONE answer from the following options:
 - Keep under regular outpatient follow-up
 - Perform a Mantoux test
 - Start on isoniazid 5 mg/kg and then perform a Mantoux test after 3 months' treatment

The Mantoux test is positive at 17 mm. The child is well with height and weight on the 75th centile and she has no fevers or night sweats. A chest X-ray shows prominent right hilum and partial collapse consolidation of the right upper lobe.

- What course of management should be followed? Chose ONE answer from the following options:
 - The child is well and should be treated for latent TB with isoniazid and rifampicin for 3 months
 - The X-ray changes are probably due to intercurrent infection and should be treated with a course of oral co-amoxycylav with a follow-up X-ray
 - The chest X-ray shows evidence of clinical disease and the child should be treated with 6 months' antituberculous treatment
- The 4-year-old had a BCG as a baby and has a visible scar on his upper arm. He is well. Which management plan should be followed for him? Chose ONE answer from the following options:
 - Perform a Mantoux test
 - No further action required as he has had a BCG
 - The child is well and should be treated for latent TB with isoniazid and rifampicin for 3 months

The child is reviewed with the results of the Mantoux test, which is 16 mm. He goes on to have a gamma-interferon test which is negative.

- What should happen now? Chose ONE answer from the following options:
 - Perform a chest X-ray and treat for active TB

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- B. Inform and advise his parents, and discharge him from the clinic
- C. Treat for latent TB and X-ray again in 6 months
- D. Perform a further BCG

Case 3

A 3-year-old boy is brought to A&E. He is with his grandmother who reports that he fell off his bike earlier in the day, but she did not think it was serious. He is now lethargic and complaining of left leg pain. On examination he is febrile at 38°C, HR of 160/min and is complaining of leg pain, but there is a full range of movement. He has two small bruises on his legs and two non-blanching petechiae on his abdomen.

1. What is the immediate management? Chose ONE answer from the following options:
 - A. Prescribe paracetamol for the pain and discharge home
 - B. Place his leg in a cast, give paracetamol and await a trauma opinion in the morning
 - C. Assess his airway patency and breathing, consider giving high flow oxygen, further assess for signs of shock, obtain IV access and give a bolus of 20 ml/kg fluid
 - D. Take a blood culture and start IV antibiotics with flucloxacillin

The rash extends and several purpura appear on his legs. The child however is much more alert and is talking normally to his grandmother. His HR remains 160–170/min and capillary refill time centrally is 4 s.

2. What is the optimal further management? Chose ONE answer from the following options:
 - A. Urgent MRI of his leg to exclude osteomyelitis
 - B. A diagnosis of meningococcal disease is likely so he should be given IV penicillin, corticosteroids and a lumbar puncture
 - C. Place on IV antibiotics and fluids and assess 2 hourly for signs of shock
 - D. Give a further bolus of 20 ml/kg of colloid, IV ceftriaxone and make contact with a PICU and anaesthetics
3. Which two of the following blood tests will be most useful:
 - A. Arterial blood gas
 - B. Clotting screen
 - C. Liver function tests
 - D. CRP
 - E. CSF culture
4. He has two brothers, aged 5 and 7, and his mother is pregnant. How should they be managed? Chose ONE answer from the following options:
 - A. Children should receive rifampicin 10 mg/kg and the mother rifampicin 600 mg
 - B. Children should receive rifampicin 10 mg/kg and the mother 500 mg ciprofloxacin as she is pregnant

- C. Children should receive rifampicin 10 mg/kg and the mother ceftriaxone 250 mg in a single IM dose

Answers**Case 1**

1. A and C

The most likely diagnoses are *Plasmodium falciparum* malaria or bacterial sepsis, which clinically can appear to be very similar, both presenting with shock. The presence of shock makes influenza less likely. That the boy has taken malaria chemoprophylaxis is no reassurance, as this cannot prevent all cases of malaria. *Plasmodium vivax* is prevalent on the Indian subcontinent and in Central America but is only rarely seen in Africa as it preferentially invades erythrocytes bearing the Duffy blood group antigen, rarely found in the African population. Worldwide *Plasmodium falciparum* is responsible for cases of severe and complicated malaria, which untreated has a significant mortality. Dengue fever is also associated with fever and shock but is much more common in the tropical areas of Asia and America and is unusual in East Africa.

2. B

Although all four of these investigations are necessary, a blood glucose is urgent in severe malaria where hypoglycaemia can occur, causing reduced conscious level and convulsions.

3. D

This child has several features indicating he is at high risk and in need of urgent supportive management. These include a depressed conscious level and evidence of shock with tachycardia and a prolonged capillary refill time. The emergency assessment and management of the child should follow the structured approach advocated in the APLS guidelines. Emergency management must not be delayed while the diagnosis of malaria is confirmed. The initial management of shock should include a bolus of 20 ml/kg of colloid or normal saline. There is no evidence that exchange transfusion has a role in the initial management of children with suspected malaria and it may distract from simpler resuscitation measures.

4. D

Parenteral quinine remains the antimalarial treatment of choice for patients with severe falciparum malaria and should be prescribed for 7 days. However, children often recover clinically before then and the course may be shortened by switching to a full oral course of an appropriate non-quinine medication. Oral quinine has a bitter taste and is associated with poor compliance; therefore, other oral medications are

recommended. Parenteral artesunate has been shown to be superior to quinine in the treatment of severe malaria in adults in South-East Asia. These results cannot however be extrapolated to children and a multicentre study in children is currently ongoing. Chloroquine resistance is now widespread and is no longer the first-line treatment for falciparum malaria.

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Case 2

1. C
2. B
3. C
4. A
5. B

The management of close contacts of individuals with active tuberculosis have to be contacted and screened and are managed according to the NICE guidelines.¹ The management is determined by the age of the child and whether they have received a BCG.

The 3 week old baby: Neonates who have been in close contact with people with sputum smear-positive TB who have not received at least two weeks' anti-tuberculosis drug treatment should be treated as follows:

- The baby should be started on isoniazid 5 mg/kg and then a Mantoux test performed after three months' treatment.
- If the Mantoux test is positive (6 mm or greater) the baby should be assessed for active TB with clinical assessment and chest X ray. If this assessment is negative, then isoniazid should be continued for a total of six months.
- If the test is negative (less than 6 mm), then isoniazid should be stopped and a BCG vaccination performed.

The 18 month old child:

BCG-vaccinated children aged older than four weeks but younger than two years, in close contact with people with sputum smear-positive respiratory TB, should have a Mantoux test. This is considered to be positive if measures 15 mm or more and the child should then be assessed for active TB. An abnormal chest X-ray in an asymptomatic child is a sign of active disease requiring six months treatment with at least three drugs for the initial two months.

The seven year old child had had prior BCG but the mantoux measured more than 15 mm so is considered to be positive. The use of the interferon-gamma blood test helps to distinguish between mantoux reactivity due to prior BCG or to actual infection with *Mycobacterium tuberculosis*.² A negative test in this child indicated the mantoux test was probably due to the prior BCG and not to either active or latent tuberculosis.

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- 2 Taylor RE, Cant AJ, Clark JE. Potential effect of NICE tuberculosis guidelines on paediatric tuberculosis screening. *Arch Dis Child* 2008; **93**: 200–203.

Case 3

1. C
2. D
3. A and B
4. C

Meningococcal disease remains an important cause of mortality in children in the UK. Studies have shown that an increased risk of death is associated with the failure to recognize complications, such as shock or raised intracranial pressure, and how ill children are; being managed by unsupervised junior doctors and by non-paediatric trained staff; and management that is often not sufficiently aggressive, as indicated by a failure to use enough inotropes in septicemic patients.¹

A key feature in this patient is the severe limb pain in the absence of any other physical signs in the limb, which is a well-established phenomenon in meningococcal disease and must not be attributed to a possible accident, especially in the presence of other clues such as fever. The pain can be very severe and children have been mistakenly put into plaster to treat presumed fractures.

Underlying disease may be very advanced by the time a rash appears. The rapidly evolving haemorrhagic rash may be a very late sign. The appearance of purpura in this boy (haemorrhagic lesions of > 2 mm) is a further sign of advancing disease. Shock is a clinical diagnosis and is clearly present in this patient. The signs are a result of circulatory failure but, as here, in early shock the child may still be alert and have a normal blood pressure.

The early signs of shock include tachycardia and a prolonged capillary refill time. If the clinical response to a bolus of 20 ml/kg of fluid is short-lived or absent, and shock does not improve or progresses, large volumes may be required (over 60 ml/kg in the first hour). In this case, there is a significant risk of pulmonary oedema, so elective tracheal intubation and mechanical ventilation should be initiated, even if there are no signs of respiratory failure, to optimize oxygenation, reduce the work of breathing and improve cardiac function. Therefore, it is important to alert senior staff early. The presence of a metabolic acidosis with a base deficit of less than –5 will give a guide to the severity of illness and the need for further fluid and bicarbonate. Coagulopathy is also a marker of severity of disease and if deranged indicates the need for fresh frozen plasma. Hypoglycaemia (< 3.3 mmol/L) is common and should be corrected.^{2,3}

All cases of meningococcal disease should be reported to public health and close contacts given prophylaxis to prevent secondary cases.

FURTHER READING

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