Ran D. Anbar
Editor

Functional Symptoms in Pediatric Disease

A Clinical Guide

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This book is dedicated to my wife, Hannah, my beloved partner and companion, who has inspired our undertaking of some of life's greatest adventures.

Preface

The effect of emotions in the practice of medicine has long been acknowledged, given their underpinning for the development of double-blinded placebo-controlled trials. Blinding of patients for research is necessary because sometimes the act of taking even an inert substance can lead to improvement in a patient's health condition (which is defined as a placebo response). This improvement can be attributed to a patient's *psychological* response to a therapy, which can lead to beneficial *physiological* changes. Blinding of the clinician also is helpful because a clinician's expectation of a therapeutic outcome can also affect the patient's response to a therapy or the clinician's interpretation of the therapeutic outcome. The importance of recognizing the significant psychological impact within medicine has been further underscored by the recent modification of the Medical College Aptitude Test (MCAT).[1] Beginning in 2015, this test will include assessment of knowledge regarding behavioral and social sciences among applicants to US medical schools. In order to broaden the knowledge of healthcare providers about the impact of psychology in the practice of pediatric medicine, this book introduces readers to the wide spectrum of functional symptoms that can affect children.

All children with medical conditions are affected by psychological factors that may have been a precursor to and/or the result of the development of their illness. Further, a significant number of these children fail to improve completely despite their physicians' best medical interventions. Sometimes, we ascribe this failure to lack of adherence to therapy or to the severity of the condition. What we often fail to appreciate or address is that the patients' psychological states may contribute to the lack of response to medical therapy. For example, some patients with asthma may experience shortness of breath as a result of anxiety rather than from physical exacerbation of their asthma. Some patients with inflammatory bowel disease may experience increased abdominal discomfort related to depression. Patients with migraine headaches may experience an increase in the frequency and severity of their symptoms as a result of psychosocial stress. Treating such patients by intensifying their medical therapy is often of little use and may even be harmful. Another group of patients have complaints that many clinicians recognize to

[1] Kaplan RM, Satterfield JM, Kington RS. Building a better physician – the case for the new MCAT. NEJM. 2012;266(14):1265–8.

have arisen as a result of functional issues such as functional abdominal pain, pseudoseizures, stress-related chest pain, and vocal cord dysfunction.

Review of the first section of this book will enable healthcare providers to recognize functional symptoms that can complicate organic disease, as well as symptoms that are believed to be mostly functional in origin. Children's healthcare providers need to keep in mind that the manifestation of functional symptoms can vary depending on the patients' developmental age. Material will be introduced through literature reviews, case studies, and quizzes. Video demonstrations are provided with some of the chapters and can be accessed on SpringerImages. Links to these videos are provided on the chapter's opening page as well as on the copyright page of the book and are freely accessible to anyone who purchases the print version.

Even when clinicians are comfortable in identifying patients with functional disorders, planning a treatment program can be a challenge. Some patients may be reluctant to undergo evaluation by mental health providers, even when such competent providers are available. For this reason, the second section of the book will help clinicians differentiate the patients for whom referral to a mental health provider is mandatory from those for whom other approaches may be useful. For the latter group, the book will teach clinicians to empower themselves by learning how to incorporate various approaches for the management of functional disorders in their practice, including acupressure, biofeedback, basic cognitive behavioral therapy techniques, hypnosis, imagery, meditation, and yoga. Means of obtaining training in these modalities are provided in the appendix.

Our target readers are practicing clinicians including pediatricians, family practitioners, pediatric subspecialists, child psychiatrists, psychologists, other mental healthcare providers, and practitioners of alternative and complementary medicine. Other readers may include students of medicine, other health and behavioral sciences, and residents and fellows undergoing advanced training.

Syracuse, NY, USA Ran D. Anbar

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Part I

Recognition of Functional Symptoms in Children

Functional Symptoms in Young People: Conceptualizations, Definitions, and Approaches

1

F. Ralph Berberich and Laurence Irwin Sugarman

Abstract

All health conditions incorporate and exhibit the interplay of psychophysiological factors, which become significant in functional disorders. Functional disorders are a subset of health conditions for which conventional medically based evaluations do not reveal structural or physiological abnormalities to account for the presenting symptoms. As such, they challenge clinicians to shift from models based on mind–body duality, and the primacy of externally applied therapies, and to work instead with the reality of psychophysiological phenomena. By recognizing functional groupings of these conditions and by developing appropriate therapeutic approaches, clinicians can help young people and their families to grow, change, and move toward well-being. In so doing, clinicians invest in their young patients' resiliency and self-efficacy, enrich and solidify patient/provider relationships, and increase clinician satisfaction in practice.

Keywords

Functional disorders • Psychophysiological • Psychosomatic • Functional pain syndromes

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Part of the joy and challenge of clinical care is that many health conditions are more complex than they first appear. Healthcare providers are conventionally taught to view patients' somatic predicaments as having developed as a consequence of measurable anatomic and physiological problems. In the absence of positive diagnostic tests, symptoms often are ascribed to psychological causes, sometimes dismissively. Then health professionals use the history, physical examination, and laboratory tests to search out evidence to support that paradigm. This approach is reinforced by the economic reality that healthcare

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systems have traditionally reimbursed providers for spending time and resources on anatomically and physiologically based diagnosis and treatment. Often set aside are the vast and powerful psychophysiological networks that comprise mind–body interactions. Less measurable, yet always at play, these interactions may be recognized but discounted, because they are neither easily addressed nor well understood. Similarly, the skills of using language therapeutically for suggestion, reframing of experiences, and behavioral change are not greatly valued, modeled, or taught in detail as part of medical training. Thus, young people who present with functional disorders provide intriguing and challenging opportunities for clinicians. The problems that exist in the mind–body realm invite physicians to break through the conventional wisdom and expand their skills in order to make a real difference for the children in their care.

This chapter presents a framework for understanding common functional pediatric problems. We briefly explore definitions and groupings of these conditions. We also stress concepts and skills that lead clinicians to embrace and therapeutically interact with the rich psychophysiological substrates available as resources to their young patients. We consider a conceptual spectrum in which purely psychological processes reside on one end and purely physiological ones on the other as a false model. It is critical to recognize that neither pole exists in reality. There is fluidity on this continuum. All physiological problems have psychological correlates as well as the reverse. The language and responsiveness of caregivers, the meaning of symptoms to patients, and the shared understanding between both are an essential part of any symptom presentation and resolution. In this lies the key to understanding functional disorders in children of any age.

Definitions, Common Notions, and Preconceptions

Functional disorders often are defined as symptoms that present without identifiable anatomic or physiologic cause. As such, they do not fit conventionally defined medical diagnoses. The term "functional" overlaps with the domains of psychosomatic conditions and conversion disorders. The expression is also used to refer to symptom responses that exceed those expected for a diagnosis and treatment pairing [1].

Some categories of functional disorders define disruption of function in an anatomical system (e.g., the gastrointestinal system) lacking an identifiable "medical" cause. These recognize psychophysiological processes, but still support a false duality. They separate what underlies the "functional" umbrella from what is said to be "organic" or "physical." For example, the definitions embodied in the Rome Criteria for functional abdominal pain are symptom based and refer to gut motility, sensitivity of the intestine's nerves, and the way in which the brain responds [2]. But by excluding conditions marked by recognized anatomic or physiological abnormalities from "functional", they fail to stress how physiology, mind, emotion, and attribution all interrelate and play off one another in the experiences of real people.

The literature is replete with evidence demonstrating that anatomic or physiological changes and psychological processes are intertwined and inseparable. For example, reports of structural brain change with irritable bowel syndrome [3] illustrate that brain/body influences are bidirectional. Even if the resolution of current medical evaluations is too insensitive to detect such aberrations, clinicians should not exclude their potential presence and significance. When viewed as bidirectional, all "medical" health conditions also are "functional." In this discussion, we focus on those conditions that presently lack evidence of structural and/or physiological abnormality, knowing that these distinctions may someday be revealed. We also recognize that clinical interventions that address functional conditions have relevance for all health problems.

Life threatening, emergency conditions excepted, it is problematic that physicians are taught systematically to "rule out" a host of detectable anatomical and physiological etiologies for a given problem before addressing psychological factors. Some clinicians may fear this could lead

them to miss a correct and important physiological diagnosis and treatment. While rational and cautious, the "medical first" approach is based on the flawed assumption that a clinician can only address body or mind, but not both simultaneously. It ignores the fact that the process of diagnostic evaluation itself produces psychophysiological effects [4, 5] which can be harmful. A medical evaluation can increase a patient's anxiety, especially when no abnormality is found and the patient is concerned that the doctor must be missing something [6]. Further investigations by the doctor would appear to support this notion, as the clinician keeps searching for a medical explanation. Finally, when a "negative" workup leads the clinician to state, "I don't find anything wrong," the implication may be left that the problem must be in the patient or parent's imagination. The phrase may also be interpreted as an expression of failure, therefore an invitation to seek "the answer" from yet another physician. At times the clinician's knowledge and authority may be challenged by the young person with functional symptoms. This can lead the clinician to feel discouraged by his or her own inability to help their patients with functional symptoms. At worst, the clinician may come to view young people with functional symptoms as malingering, attention seeking, or consciously attempting to use factitious symptoms. Interestingly, clinicians who have not chosen a mental health specialty are not taught to do the reverse, namely to "rule in" a host of psychological factors while simultaneously exploring other aspects of presenting symptoms.

We therefore wish to emphasize that physiological processes (e.g., inflammation, muscle fatigue, and tissue injury) have neuropsychological effects (see Chap. 8) and psychological processes (emotional stress, guilt, and learning) have physiological concomitants. A person with a given presenting physical problem is also coping, at least unconsciously, with the psychological factors that exist in combination with it. Psychophysiological processes must be "ruled in" in a purposeful manner to obtain and share a more complete understanding of the young person's health conditions. So we can wonder: What if parents and clinicians all recognized and taught children that the "mind is also in the body"? Would children change how they present their symptoms and doctors evaluate those symptoms differently? For example, we have encountered children who present for immunizations prepared to "blow away" needle pain by virtue of observing their older siblings learn the same skills. Also, we have seen children who have previously learned to apply skills in relaxation and self-hypnosis return to us in order to discuss how they can apply their skills to treat other discomforts such as recurrent abdominal pain. If the ultimate diagnosis is appendicitis or Crohn's disease, these skills will be valuable adjuncts to surgical and medical care. If the diagnosis is irritable bowel syndrome, these skills may be central to the treatment plan.

Patients' and clinicians' reliance on external therapies has consequences as well. That cultural emphasis, currently best exemplified by direct-to-the-consumer advertising of prescription medications in the USA, may increase both somatization and the promotion of physiological and pharmacological treatment of psychological symptoms [7, 8]. Patient and doctor alike are primed to attribute an anatomic or physiological cause to most conditions or, if not, attribute them to a solely psychological or social basis. Young patients and parents also tend to assume that their symptoms are the result of external forces rather than recognize that they may be a product of their own unconscious processes. This is evident from placebo research which demonstrates that conditioning and expectation associated with inert interventions, whether subjects are blinded or not, can be associated with significant neurophysiological effects that are as effective as pharmacological or comparably "active" interventions [9].

The tendency to attribute the trigger of our symptoms to external causes is also revealed in the language commonly used to describe functional symptoms. For example, a child with a stress-induced headache is likely to say, "Doing homework after school, I *got* a headache *that would not stop*. I had to go to bed and take a nap. Then it *just went away*." The italicized words indicate the child's passivity and externalization. A more accurate description could be, "Because it is so

important to finish my difficult homework, I *wouldn't let myself stop* doing it until *my unconscious allowed me* to *develop a headache* so I had a good reason to stop. *Then I gave myself* a nap so *I could turn down the pain*, even though I really needed it to get away from the homework and rest." Here, the italics denote active, child-enacted behavior. In short, we are conditioned to ignore our own innate abilities and instead ascribe them to external interventions. The long-standing emphasis on medical care of the physical body has only served to augment and maintain the dominance of that perspective.

In order to consider, evaluate, and intervene effectively, clinicians need first to shift from the false dualities to models of health and disease that include bidirectional psychophysiological currents of information. By learning to evoke each individual child's innate creative abilities and talents for helping him or herself in concert with external interventions, clinicians can allow their young patients to balance pills with skills. Imagine walking into the examination room to see a child in pain and initiating the encounter by saying, "You look pretty uncomfortable. I bet you would rather be someplace more comfortable. I wonder if going there in your mind's imagination right now will help you feel better while we figure out how else to help you." As the child's eyes shift away from you, you add, "That's right. Just like that…"

Clinical Vignette

The following vignette illustrates the approaches discussed in the rest of this chapter. Italics are added to emphasize the therapeutic language that is critical to helping children with functional symptoms tap into their inner resources when confronted with those symptoms. This example also illustrates what so frequently confronts a busy, time-pressured clinician: the expectation is that the care provider's role is to diagnose and treat a physical illness. The notion that a symptom is only partially or not at all due to recognizable physical illness may be met with skepticism, resistance, or hostility. But, as we discuss, a symptom represents the individual's best effort to mount an adaptive, beneficial response to a problem. That response may be either beneficial or maladaptive.

EJ was a 17-year-old girl who was new to the primary care clinician's practice. She presented with 5 days of throat pain and impaired swallowing by self-report. She was hungry, but felt she could not eat due to pain. She stated that she had a low-grade temperature at the onset of symptoms, but that it resolved on the second day. She felt well otherwise. When asked to describe the sensation, she said she had "a lump in my throat." On examination she was serious, focused on her symptom, and concerned, as was her mother who was also in the room. EJ's vital signs and physical examination were within normal limits. Specifically, she had no evidence of pharyngeal inflammation, or lymph node tenderness. Her gag and swallowing reflexes were normal except for her apprehension during the pharyngeal exam. She winced when swallowing. Her voice was quiet, tense, and slightly coarse as EJ asked anxiously, "What do I have?"

While an inflammatory process was part of the differential diagnosis, the absence of evidence for pharyngitis and associated systemic symptoms makes the diagnosis of a functional "globus" reaction more likely. The clinician told the girl that he planned to rule out a streptococcal infection and then softly, gently and kindly empathized, "These lumps in the throat are really stressful, aren't they?" She nodded and wiped away a tear. He gently swabbed her posterior pharynx to test for streptococcal antigens. Returning while the test was in process, he asked the girl what other stressors she might be coping with. She responded, "Well, there are my college applications…I don't like deadlines." He reflected, validating her statement, "College applications are a big deal." As she began talking about her choices, dilemmas, and aspirations, her voice loosened and became clearer. Glancing at her mother, she explained that she wanted to base her decision as much on the living environment as the academic reputation. He commented, "That can be a wise thing to do," because "When a person is happy in her new environment, she *finds it easier* to study and learn and *find success* both socially and academically." She became more animated as

she went on to discuss her potential college choices. He noted that her affective range broadened and her voice became more melodic as the tension left her speech. With this apparent release of tension, he asked whether there were additional stressors. She quieted, then again became tearful, and explained that her mother was awaiting results of a skin biopsy to "rule out cancer" adding that her maternal grandfather had recently died from a form of skin cancer. The clinician again commented empathetically, saying, "That, would seem to be a *lot to swallow* as you make your plans to *move into your life*." Then he left the room to retrieve the test results.

The rapid antigen test was negative. On his return, EJ asked what would make her throat better. The clinician asked her what she thought would be of most help, noting that her voice, "already seems more *comfortable and strong*." Without prompting or direct suggestions on the part of the doctor, EJ replied, "I think it will get better all by itself." He replied, "Well I am not surprised that you would say that. I already told you that I thought you were a pretty wise young woman. But I do wonder whether that throat of yours really is getting better "all by itself". I think that *you are* actually *making your throat more comfortable and relaxed*." He also mentioned that symptoms are the body's way of telling us "We *need to change* what we are doing," in her case by tightening up and closing off to any more "stuff to deal with." She added that "talking about things must help my throat to settle down." Nodding, he wondered what first "*cool, soothing*" drink she would choose to talk her mother into getting her after she left. She chose a chocolate milk shake at a favorite fast food restaurant. He suggested that she might as well begin to "*Taste it and feel it* now. Why wait? It can't hurt." He urged she return to school so she "could *start feeling normal*" but also to "*take everything easy*." Finally, he asked her to make an appointment for the following week, "Just to talk about how well you are handling everything." He explained that she could cancel it only if she left him a message about how well she was doing.

This vignette emphasizes an interactive therapeutic approach to eliciting the history of patient's experiences with her illness [10]. In this case, the clinician expanded the scope of the presenting problem to include the stressors that EJ offered as relevant, and then validated her priorities and choices to support her self-efficacy. The clinician also allowed his absence, while retrieving test results, to let her consider the significance of this framing. He carefully crafted his language to intersperse empowering direct and indirect messages about the resolution of the symptom. His empathy with EJ and interest in her well-being beyond the presenting symptom supported rapport, evidenced by her willingness to further reveal her concerns with the clinician whom she had just met in an acute care environment. He explicitly expressed his confidence and encouraged hers by asking her what she thought would help her, thereby further evoking her own resources. Recognizing that a very brief visit might not provide sufficient support, he requested a follow-up visit framed neutrally with the trust that she could opt out due to improvement.

Functional Groupings

In order to understand and be able to compare different research results, categorization is necessary, despite being a contrived and imperfect fit for an individual patient. It therefore is useful to consider distinct symptom groups, identifying those conditions with primarily physical symptoms and those with primarily psychological origins. Common pediatric functional somatic states can be grouped into three broad categories, pain syndromes, cardiorespiratory syndromes, and chronic fatigue/weakness. Primary manifestations of pain, headache, and abdominal discomfort are common, while musculoskeletal syndromes are less frequent in general, and mostly affect adolescents when they occur.

Anxiety-driven conditions and habit disorders are primarily functional in origin and include tension headaches, school or social phobia presenting as abdominal pain, trichotillomania, habit cough, globus, hyperventilation, nail biting injuries, and neurodermatitis. The same applies to fatigue and insomnia. Furthermore, each of these functional entities can be associated with other diagnoses as a consequent or concomitant factor.

For example, anorexia nervosa can have profound anatomic and physiological consequences along with functional ramifications well beyond food aversion and the preoccupation with its restriction.

Incidence/Prevalence of Functional Complaints

Consistent incidence and prevalence rates of functional complaints and disorders are hard to come by due to varying classification of somatic and psychophysiological symptoms and conditions, ethnic and national differences, and different age groupings analyzed. Most prevalence studies suggest that 20–25 % of children experience at least one somatic symptom, with lower incidences in early childhood that rise into adolescence [11, 12]. An elegant recent review by Campo lists a number of studies that address incidence and prevalence particulars [13]: Over the course of development into adolescence, female preponderance increases. Any single functional symptom tends to be associated with others up to 40 % of the time, and the clustering of functional symptoms has been reported as at least four different manifestations in 12–15 %. Abdominal pain predominates in preschool children. Headache and abdominal pain are the most common functional symptoms in school age children, reported on a weekly basis by approximately 10–30 % and 5–7 %, respectively. Headache and musculoskeletal complaints become more common in older age groups. In teenagers, somatic complaints are encountered at an approximate rate of 10–15 % of girls and 4.5–10 % in boys [14]. Teenagers frequently report fatigue, weekly in up to half, and daily in 15 % [15].

Correlates of Functional Disorders

Certain functional symptoms tend to accompany each other, even when the precise origin and sequence of their onset and progression are muddled. For example, functional abdominal pain, anxiety, and depression often are cooccurring. Depression is less common when abdominal symptoms are associated with structural or physiological abnormalities [13]. Generalized pain, unexplained pain in different locations, and multiple somatic complaints both are associated with depression and anxiety. Similarly, when depression exists, somatic complaints are relatively more likely to arise [16, 17]. All three conditions tend to overlap with varying temporal manifestations. All tend to respond similarly to cognitive behavioral and psychopharmacological treatment [13, 18], and the conjunction of functional somatic disorders, anxiety, and depression appear to be constant across the lifespan [13].

Two other correlates of note are the meaning-importance and the consequences that derive from a functional symptom. Children with chronic physical ailments may develop amplification of sensory stimuli, especially when their attention is captured by the identification of a specific problem area [14, 19]. This increase in sensitivity can provoke what seems like disproportionate reporting of symptoms. For example, a child who has chronic asthma, with its associated symptoms related to the lungs, may become increasingly sensitive to the sensation of shortness of breath, especially if he has experienced a frightening episode. This can result in his reporting, “My wheezing is worse,” when objective measures such as a pulse oximetry reading or pulmonary function tests are unchanged. Another source of tension, such as a school test, may express itself as a chest symptom.

Symptoms may increase parental attention or represent a mechanism to avoid an object of fear or conflict [20]. Social factors in the extended family, from school or among peers, may reinforce the consciously or unconsciously perceived benefit of somatic symptoms. Terms such as “primary and secondary gain” have been used in the past to describe these patterns. Strategically, we suggest that the use of these terms run the risk, at least in the clinician’s mind, of implying that the young person is using these physical symptoms purposely to avoid something that causes him even more psychic pain, perhaps even malingering. It is critical that such an implication not make its way into the clinical interaction, because rapport and trust are undermined by disbelief and demeaning pronouncements

("That can't be true = You are a liar"). This in turn inhibits effective diagnosis and treatment.

Temperamentally sensitive and insecure, conscientious young people, who strive for high academic achievement, tend to somaticize [14]. Perhaps, this is because the burden of perfection is unsustainable. Thus, functional symptoms can be viewed as adaptive when they autonomously "solve" an underlying problem. For example, consider the child who develops abdominal pain, nausea, and vomiting on school mornings, which provide an excuse for avoidance of a stressful situation at school. At the same time, functional symptoms also are maladaptive when they conflict with vital childhood activities and pleasures, such as education, sports, social life, friendships, social development, or maintaining appropriate nutrition or hygiene.

Family and Social Contributions

Children participate as members of a family system. The composition, proclivities, and success of functioning within that family often play a crucial role in the generation of functional symptoms. Parental demeanor projecting excessive worry and a sense of catastrophe often will cause exaggerated responses in children [20]. Somatization and poor self-regulatory skills, impaired judgment, and diminished impulse control often coexist intergenerationally and so resonate within families. The resulting discord or dysfunction within the family may trigger a child's anger or repression along with functional somatic expressions of distress. This may also contribute to functional disorders because there is evidence from a community-based study documenting that children with inadequate emotional awareness skills and negative affect increased their self-reported somatic symptoms and sympathetic arousal [21]. The same study linked parental somatization with somatization in their children, perhaps through modeling, prompting, or increased attention to somatic complaints [21]. As children grow older, they may accept and resonate with parental beliefs and attitudes about the presenting symptom. Thus, family dynamics and established patterns of functioning may arouse conflicts in a young person at various times leading to functional disorders.

Most functional problems come to the attention of a clinician because of persisting pain, a visible or perceived change in the body, school absenteeism, sleep issues, or disruption of some aspect of family life. Every one of these elements has the potential to worsen what ails the child, creating a spiral of increasing and persisting pain or dysfunction [22] and a loss of self-esteem and self-efficacy. It may be necessary to dissect the social fabric enveloping the child to understand why a functional symptom has arisen.

To complicate matters further, some parents anticipating a "medical" diagnosis may have a set belief that rejects any possibility of a psychological origin of the symptom or emotion-driven contributors. This conviction can derive from their own upbringing, the concern that any emotional problem reflects badly on their parenting, or, more likely, both. In this circumstance, the practitioner must pay particular attention to his or her language and the responses of the child and parent. The subject should be introduced in a way that allows for understanding as part of the interactions while the history is obtained from the patient and family. Dismissive language and gestures can create resistance to the notion of a functional etiology. A useful strategy is to ask the child and parent what they imagine and fear might be causing the symptoms. In the authors' experience, given a receptive listener, young people and their parents more often bring up mind–body phenomena, than reveal their ignorance of the same. The integration of mind–body relationships throughout the continuity of care and as part of anticipatory guidance and the methodology that allows it to emerge into the open are exhaustively discussed in texts and other published materials [23, 24].

The Clinician's Role

Establishing a Functional Diagnosis

The clinician who treats children is accustomed to mystery and ambiguity when seeking to unravel etiologies of symptoms. Depending on age and stage of development, young patients

will have varying ability to disclose symptoms and may deliver a partial history or none at all. Then, the parent becomes the "interpreter," for better or worse. Additionally, challenging is that the clinician's office itself can become an unpleasant, hostile, feared place for the child, and identified with discomfort, pain, and coercion. Children so affected already have a fear of the doctor that can cause distortion of history, signs, symptoms, and the physical examination. The medical encounter also may be defined by fear of injections [25–27]. The child who dreads the needle, otoscope, or throat swab might deny symptoms reported by the parent to avoid an injection, ear exam, or throat culture.

Establishing a functional diagnosis requires skill and patience. A functional component or origin must be included in almost all differential diagnoses. Some parents may be dismissive, seeing these problems as "not medical" or not taken seriously. When discussing circumstances that might expose them, some children may complain, "I thought this was for a checkup?" When discussing a symptom or diagnosis, the clinician should explain that exhaustive laboratory testing and imaging "for completeness" need not be performed for every presentation. Depending on the risk attached to a rare and unlikely possibility (e.g., brain tumor causing a headache), treatment for a diagnosed functional condition can and should take place concurrently, even while remaining open to the possibility that a "medical" disorder has not yet been uncovered. Because no laboratory or imaging test is available for a functional diagnosis, some uncertainty and fluidity must be accepted in partnership with the family. For example, an otherwise well child with a normal physical examination struggling with academic performance and frequent headaches on nights prior to school can be presumed to have functional headaches. That is a valid preliminary diagnosis and treatment can and should be directed toward addressing it. Because differential diagnosis is an ongoing undertaking, it continues, until a problem is resolved completely. Less likely possibilities should be pursued if a change in the clinical course warrants. So, if this child were to develop daily early morning vomiting and weekend headaches, further evaluation would become urgent.

The patient–parent interview can open or close the opportunities for functional diagnoses. Direct confrontation usually does little more than provoke resistance. Trust and rapport are vital. Language is important and one must choose one's words carefully. Questions phrased as "How….?" Or "What….?" followed by open-ended phrases such as "Tell me more about…" or "I wonder when…" are more likely to allow patient and parent to speak openly. One can appreciate the differences between "Do you get nervous over every test?" versus "How do you feel when you have a test? What does it feel like? Where in your body do you feel it?" Sometimes a parent feels a strong, protective need take over the interview and the child's own narrative is lost. The clinician is urged to solicit the child's own comments while also validating the importance of the parent's presence and contribution. Nonverbal responses of the young person and his or her parent can be as important as the information conveyed through speech. When a child says that his abdominal pain feels like "my stomach is fighting and coming apart," the clinician might then explore the possibility of a metaphoric linkage. Thus, "my stomach…. fighting and coming apart…" may refer to a recent marital separation, an ill relative, or other perceived or experienced rifts in the child's social fabric. Academic performance, social stress, other sources of anxiety, and level of self-esteem are important areas for careful probing. The doctor should touch on bowel function, sleep patterns, and emotional factors, in the review of systems when evaluating most symptoms, and any that suggest functional possibilities. Verbal or nonverbal expressions of embarrassment at discussing "personal" matters should be greeted with demystification, understanding, and encouragement, so as to allow their inclusion. It may help to frame the approach by saying, "So many other young people I see with stomach aches also struggle with [family problems, sibling dynamics, school, etc.]. Does that happen to you?"

The long-term continuity of care that primary care physicians enjoy allows for the integration

of the past history, the particularities of a child's developmental trajectory, and preexisting conditions of temperament or learning style to enter into the diagnostic potpourri when confronted with a new symptom. These relationships also create opportunities for ongoing incremental teaching about functional aspects of symptoms and the promotion of psychophysiological adaptability in each child and family.

What might hinder the clinician from identifying a functional disorder? The diligent physician is ethically and efficiently obligated to make the correct diagnosis and avoid significant errors of omission. In some contexts, the fear of litigation can drive testing in pursuit of an objectively quantifiable diagnosis. Failure to diagnose a significant and treatable medical illness risks monetary penalty as well as professional and personal censure. Quality and standard of care concerns can promote testing at the expense of a timely functional diagnosis. There may be physician reactivity, an assumption that a patient presenting with symptom always calls upon the doctor to "do something" involving medication or a procedure. There is some danger that relentless pursuit of a defined "medical" cause or "pathologizing" a symptom can lead to iatrogenic harm [28]. The data collection necessitated by today's electronic health records may interfere with the totality of observation if the doctor does not know how to type without looking at the keyboard [29].

Anticipatory Guidance to Promote Mind–Body Phenomena

The clinician can use regular well child visits to teach the notion of mind–body interdependence. That process aptly begins during infancy when the primary care physician utilizes discussions about developmental milestones to help parents foster adaptation and resilience in their child. The clinician can note, in lay terms, how the child is creating his or her own unique neurophysiological networks that connect emotion and attachment with expanding motor, sensory, and language abilities. For example, when the infant or toddler takes her first steps and the proud parent relates this to the doctor, the clinician can say, "Have you noticed how, as Anna masters her walking in her own way, she checks to see if you are looking when she falls, then decides whether to cry or just get up and keep going? Yes. All of us have done that, and it can be hard to know how to react. But she learns the most when she just keeps getting back up. She keeps figuring it out in her own way. She gets a new perspective when she gets up and walks, doesn't she?" From the newborn period onwards, the clinician can talk about common phenomena such as crying, tantrums, the resolution of infections and healing wounds in terms of mind–body reactions and primordial protective mechanisms, individual differences, brain-initiated modulation, and social and family circumstances.

Clinical encounters that entail some discomfort afford another powerful opportunity to illustrate mind–body interactions to the child and family. The physician is in a position to provide measures that encourage self-regulation, and mastery and resilience in the child. Children taught to "blow away" injection pain and fear "so you can keep it from bothering you" learn that conscious use of their breathing and focusing can change their perception of pain and diminish their anxiety. Similar strategies can be used when swabbing the throat, looking in the ear, palpating the abdomen, tending to wounds, and manipulating the injured limb. This is explicitly not about distracting the child with an external stimulus. Telling a child that they can help themselves by blowing on a pinwheel or studying the content of a picture is empowering and helps them redirect their own attention. Conversely, telling a child to "look at that funny picture over there" is an attempt to externally distract their attention. Internally-generated solutions are about evoking and generalizing innate abilities for comfort and coping [23, 24]. Over time, such experiences broaden the young person's awareness of mind–body interactions and capacities for change.

The clinician can point out that many adults no longer recall or apply how the world appears to children, because adult minds are different from that of their children and relate age appropriately to their bodies. Parents may impose adult

values, conscious thought processes, mores and expectations in a manner that does not fit the normal abilities or proclivity of the child. This may cause conflict and manifest as functional symptoms. Parents may not even appreciate the primacy of their own subconscious thought and motivations. That can lead to disappointment or inappropriate coercion, which in turn has an impact on a child's mind–body responses. A 5-year old who is scolded for not making his own bed by himself "to learn how to be neat" may "know" better than his parents that the expectation is not something he can achieve at his age. Unable, or too frightened, to object, he wets the bed. That results in further scolding, and he subsequently develops abdominal pain, which the parent brings to the doctor as a "medical" problem.

Clinicians can teach parents that children's subjective feelings, such as pain, itching, nausea, fatigue, too hot, and too cold, are recognized and processed by their brains; they are as real to the brain as things we see, hear, or touch. Feelings and sensations and "objective" findings are equally legitimate in general and particularly according to the age and stage of the child's development. Parents can be taught neither to ignore, berate nor excessively mollify a child for having physical complaints, functional or not. Even presentations that appear to represent intentional malingering deserve respectful attention and treatment.

Training and Skills

Clinicians who wish to help young people with functional symptoms require experiential training in the strategic use of therapeutic language and responsiveness to patients' cues. While training in cognitive–behavioral therapy (Chap. 19) provides some content and process, the authors' experience is that professional training in hypnosis with children gets at the core skills needed (Chap. 21). Clinical hypnosis is far from the portrayal by lay media, stage hypnotists, and traditional cultural portrayal of hypnosis as the Svengali-like external application of undue social influence. Clinical hypnosis with children rather evokes the natural resourcefulness of young people to ethically and empathically invest in self-efficacy, mastery and, critically, psychophysiological self-regulation that can be applied both for treatment and prevention. Biofeedback training (Chap. 20) is an extremely useful complement to the strategic use of language, responsiveness to states of awareness, and the building of rapport that is inherent in hypnosis [30]. Biofeedback provides both operant conditioning and cognitive training in psychophysiological regulation by simply showing the user the physiological signals they generate and then allowing them to control them in a desired direction. Biofeedback then is simply a physiological mirror. It both demonstrates the psychobiological link and reinforces self-regulation. Biofeedback training can be performed with equipment that is already in pediatric medical settings: a pulse oximeter, a stethoscope, or an indoor–outdoor thermometer (for skin temperature). Integration of other, relatively simple skills, such as progressive relaxation and abdominal breathing, into practice can help young people recognize that they have innate and powerful capacities for adaptive self-regulation.

Brief Vignettes and Questions

The following clinical settings are provided to stimulate the reader's creative thinking and discussion in applying concepts described in this chapter. For each vignette, consider the following three questions:

Questions

1. What are your immediate impressions of the etiologies of this young person's problems?
2. How might you proceed to help him or her?
3. Given his or her story, what might have been done earlier in life to decrease the current symptoms?

Vignette 1

A 10-year-old boy presents with a "scratchy throat" and concern that he has "allergy reactions." His mother explains that he had an anaphylactic reaction to peanut at age three with tree-nut and cow's milk allergy diagnosed by an allergist subsequently and no significant allergic reactions since. Over the last 2 years, he has greatly restricted his diet and refused to eat at restaurants or other people's homes. He brings his own lunch to the cafeteria at school. He has been refusing to go to school, complaining of a scratchy throat that "feels like it is closing up" since a student ate a peanut butter sandwich near him during lunch 3 days ago. When asked as part of the history taking, his mother explains that she has an anxiety disorder.

Vignette 2

A 14-year-old girl presents with a 2-month history of recurrent abdominal pain, early satiety, changing stool pattern (constipation alternating with loose, frequent stools), but without melena, hematochezia, nausea, vomiting, weight loss, or other systemic symptoms. There is a history of recurrent abdominal pain associated with transitions in her life over the last 6 years. She states that she has been told that it is "just stress and will go away." This episode is the longest lasting and most severe. She moved to town with her family last summer and started a new school 3 months ago. There is a family history of Crohn's disease in a second-degree relative.

Vignette 3

A 7-year-old boy is brought in by both of his parents with a 3-month history of increasing sensory sensitivity to clothes that has resulted in tantrums, school tardiness, and increasing stress in the family. He was diagnosed with Juvenile Rheumatoid Arthritis 2 years ago that has been difficult to manage. His father explains that when he was first diagnosed he was hospitalized and traumatized during an arthrocentesis of his knee. Thereafter, he has consistently told the pediatric rheumatologist that he feels fine and has no problems. At home, he complains that he "feels bad everywhere" most mornings, especially when he puts his clothes on for school.

Conclusion

Young people with functional conditions are coping with a rich interplay of psychological and physiological factors. In a real sense, children and adolescents develop functional disorders because they need to better learn and cope with how their minds and bodies are interconnected. Thus, the presence of functional symptoms provides clinicians with the opportunity, to help young people with this learning. In order to meet that challenge, healthcare providers must maintain the perspective that all health conditions include psychophysiological factors. Clinicians who cultivate the ability to simultaneously evaluate structural and physiological etiologies while exploring and addressing salient psychological factors not only are more likely to create efficiency and improve health outcomes, but they also invest in their young patients' resiliency and self-awareness. Therein lies the satisfaction of helping young people grow well.

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Functional Neurological Disorders: It Is All in the Head

2

Linda Thomson

Abstract

The stressful events in a child's life may not always be expressed in words, but rather in physical symptoms. This form of "body language" may be an alternative method of communicating stress. Psychological factors are frequently the cause of functional neurological disorders such as headaches, psychogenic nonepileptic seizures, motor conversion disorders, and tic disorders. Organic or pathophysiologic causes for the physical symptoms must always be explored. However, failure to recognize the impact of psychological factors on the symptoms may delay effective treatment and expose the child to unnecessary medications, diagnoses, and labeling, with potentially untoward side effects. There may also be significant costs for extensive laboratory testing, imaging studies, and referrals that may not be required when an accurate diagnosis of a functional neurological problem has been established. Treatment strategies for somatoform neurological symptoms are often multimodal and quite different from neurological problems with a physiological cause.

Keywords

Psychogenic neurological disorders • Nonepileptic seizures • Psychogenic seizures • Pseudoseizures • Conversion disorder • Tic disorder • Tourette syndrome • Stress headaches • Psychosomatic headaches

Background: Literature Review

There are several theories that have been proposed to explain the onset and continued expression of psychogenic neurological symptoms. One explanation is that the threatening emotions are repressed or suppressed inwardly and expressed symbolically through physical symptoms. The psychosomatic symptoms become a metaphor for the interpersonal conflicts and emotions experienced by the child [1].

Another view is that the child learns a "sick role" that is reinforced by the attention he receives from family and friends. The child may have unconsciously learned to imitate the symptoms of another.

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It is not surprising for a child of a parent with migraines to complain of headaches when stressed or desiring to avoid an activity. The physical symptom serves an important function and gain for the child. It provides an acceptable excuse for avoidance. The avoidance in turn reduces the child's anxiety. This view of the social learning theory is based on modeling and social reinforcement [2].

A third proposed explanation for psychosomatic neurological disorders is known as the stress-coping model [3]. This is conceptualized as what happens physiologically and psychologically when the demands placed on the individual exceeds the child's resources to cope. The child's coping mechanisms are overwhelmed and insufficient to successfully manage the stressful situations of his life.

The interaction of mind, body, and spirit underlie psychogenic disorders. This integration of the physiological, psychological, and sociocultural factors that impact somatic symptoms is known as the biopsychosocial model. The stressors that precipitate, maintain, or aggravate somatic complaints may be environmental, physical, or emotional [4].

Headaches

Headache is a common condition among children and adolescents and can result in considerable distress, pain, and functional disability. The prevalence of migraine headache has reported to vary from as low as 1.2 % in the preschool years to as high as 23 % among high school adolescents [5].

A Finnish study matched 96 children with headaches to controls. Migraine headaches were diagnosed in 58; the remaining 38 had tension-type headaches. The researchers found that children with headaches were more often extremely sensitive to pain. These children were more likely to become stressed with physical examinations, immunizations, and blood sampling than the controls. The mothers of children with tension-type headaches reported more sensitivity to pain than the mothers of children with migraine headaches. In this study, children with tension headaches had a more stressful family environment than children with migraines [6].

In a study by Ekstrand et al., the presence of psychiatric disorders reduced the likelihood of neurological disease among neurology referrals, particularly those with headaches [7]. This would suggest that when individuals with psychiatric issues have neurological symptoms that a psychosomatic cause should be considered. Patients with recurrent pain without organic etiology reported significantly higher life stress than patients with organic findings [8].

In a French study, children were asked to draw a picture of their headaches. When children had tension headaches rather than migraines, their drawings demonstrated more squeezing, tightness, and compression. Fifty-seven percent of the children who had headaches diagnosed as "other (than migraine or tension)" had somatoform disorders [9].

Chronic daily headache (CDH) is a term used to describe when a child has a headache for at least 15 days/month for over 3 months without any underlying organic pathology. This has been shown to represent 60 % of cases in pediatric specialty clinics. Young people frequently have comorbid symptoms including dizziness, sleep disturbance, fatigue, problems with concentration, anxiety, frustration, and sad mood. Additional pain symptoms such as abdominal, neck, back, and diffuse muscle and joint pain also may be expressed [10].

Psychosocial factors are not only the leading cause of headache in children when there is no organic pathology but also can significantly increase the frequency and intensity of symptoms when there is a physical cause for the headaches. There may be issues relating to school such as bullying, social isolation, learning disabilities, or pressure to excel. Family conflicts, child abuse, problems with personal relationships, grief, and loss may contribute to symptoms and complicate management. Drug and alcohol use not only by the child but also by the family must also be explored. Sleep and eating disorders should be considered.

When the cause of functional symptoms is psychosocial, analgesics are most often ineffective. Overuse of over-the-counter analgesics have a high potential for rebound and should be avoided.

The cause of headaches perhaps most feared by parent and child alike is increased intracranial pressure from a space occupying lesion. The comprehensive medical examination for the child with headache must include examination of the skull, brain, sinuses, teeth, eyes, ears, and cervical spine. Intracranial pressure, temporomandibular joint, cranial nerves, along with the supraorbital and occipital nerves must be evaluated [3, 11].

Nonepileptic Seizures

Nonepileptic seizures (NES) are paroxysmal behavioral events or disturbances in consciousness that resemble epileptic seizures (ES) but are not caused by epileptiform discharges in the brain. Because NES resemble ES, children are frequently misdiagnosed and inappropriately treated resulting in significant morbidity. NES are common: 10–20 % of children referred to epilepsy centers actually have NES [12].

The largest group of patients with NES have psychogenic seizures. Tonic or clonic movements, tremors, twitching, shaking, unusual postures, altered emotions and sensations, disturbances in consciousness, syncope, eye flickering, vocalizations, myoclonic jerking, or pelvic thrusting may be a response to a variety of emotional stresses. The child has no conscious awareness of the motivation behind this unique behavioral event [13, 14].

Although the NES seizures are involuntary, the seizures often serve a purpose. The seizures may allow the child to escape something unpleasant. The behavior may evoke sympathy from the parent or teacher or make the child feel more special than a sibling. Some psychogenic seizures are a manifestation of posttraumatic stress disorder (PTSD) and may represent a defense mechanism to handle physical, sexual, or emotional abuse. Some psychogenic seizures represent a conversion disorder or a behavioral (e.g., autistic) disorder rather than a real seizure. In a conversion disorder (see below), psychological stress is expressed as a physical disorder with family dynamics a frequent contributing factor. The motor movements are caused by subconscious processes responding to psychological conflict.

Another type of NES is termed pseudoseizure, which comes from the Greek meaning of false. These seizures are a manifestation of malingering. The seizures are intentionally faked for secondary gain. Due to the seizure, the child may get out of doing something he prefers to avoid or get more attention from friends or family. In adults, financial gain from a law suit or obtaining disability benefits may be the motivating factor. The child is well aware of the intention, motivation, and secondary gain from the seizures [15].

A study by Wyllie et al. examined the psychiatric features of children and adolescents with NES. The study did not distinguish between psychogenic and pseudoseizures. They concluded that major mood disorders and severe environmental stress, especially sexual abuse, are common among children and adolescents with NES. There was a subgroup of children with NES who had less severe psychiatric problems and moderate psychosocial stressors [16]. Pakalnis et al. looked at the psychiatric and other risk factors of children who had repetitive psychogenic seizures severe enough to mimic status epilepticus. All episodes of nonepileptic status epilepticus were preceded by acutely stressful situations. Anxiety and affective disorders were the most common comorbid psychiatric diagnosis [17].

An absence of relevant psychological factors was found in only 5 % of the 185 patients with NES studied by Moore and Baker. The most common psychological factors associated with NES in this study include: anxiety or stress, physical abuse, significant bereavement, family dysfunctioning, relationship problems, depression, and sexual abuse [18].

The major difficulty in making a correct diagnosis is distinguishing psychosomatic illness from seizures with an organic etiology. In a study of 43 children and adolescents with NES, nine were found to have an abnormal neurological past history. There was a family history of epilepsy in 34.9 % and often children have watched other family members' seizures. Neuropsychological testing done on 22 cases failed to show major abnormalities. Most cases, however, demonstrated significant personal and family distress [19].

There are historical and clinical clues that should help the clinician distinguish between NES and ES. NES may be suspected when there is a history of psychiatric illness, panic attacks, PTSD, suicidal ideation, and depression. Organic cerebral dysfunction following a brain injury may result in compromised adaptive abilities and subsequent onset of NES. A history of physical or sexual abuse may also increase the suspicion of NES. Antiepileptic drugs (AEDs) can facilitate NES. Therefore, if a child with ES has an increase in seizure frequency or a change in the seizure type when a new AED has been instituted despite therapeutic levels, NES should be considered in addition to ES. Due to the exquisite sensitivity of the temporolimbic structures, especially the amygdala, to hormonal balance, the onset of menarche during adolescence can influence NES [13].

Some of the distinguishing clinical characteristics of NES include longer duration with gradual onset, and dramatic movements associated with unresponsiveness but without clear loss of consciousness. NES tend to have out of phase motor activity without incontinence, injury, or postictal confusion. Hyperventilation and weeping may occur with NES. On EEG, there will be muscle artifact, but no ictal build-up or postictal slowing. Unlike ES, there will be no rise in prolactin postictally [13, 14].

An added diagnostic conundrum is frontal lobe epilepsy, which frequently presents in the first or second decade of life with no abnormal EEG or radiographic findings. Thus, identifying the key clinical characteristics of frontal lobe seizures is probably the best diagnostic tool. The motor activity may be frenetic, semipurposeful movement, pelvic thrusting, or tonic–clonic. The patient having a frontal lobe seizure may yell, grunt, or shout obscenities. The level of consciousness may range from full conscious awareness to complete loss of consciousness with rapid return to baseline. Frontal lobe epilepsy usually occurs while the patient is sleeping, may be associated with leg restlessness, and often occurs in clusters. There is an increased risk of status epilepticus with frontal lobe epilepsy, so an accurate diagnosis is important [20, 21]. Complicating the diagnosis even further, an Indian study suggests that children with ES are more likely to manifest NES [22].

The gold standard for diagnosing NES is continuous video EEG monitoring with close circuit television to compare and contrast the clinical and EEG features of the videotaped events. A single normal EEG tracing cannot rule out seizures. An MRI, Positron Emission Tomography (PET), Single-Photon Emission Computed Tomography (SPECT), psychological evaluation, and neuropsychological testing are all part of the evaluation.

Conversion Disorders

When there is an alteration or loss of physical functioning that appears to be a physical disorder in the absence of an organic etiology, a conversion disorder is highly probable. For example, a child may report that he is unable to see after viewing something horrific. The conversion reaction acts as a protective defense mechanism. The sensory and motor dysfunctions can encompass any nervous system activity that is to some degree under voluntary control and may be an expression of some psychological need or conflict [23, 24]. The symptoms may present as motor paralysis, weakness, blindness, NES, swallowing difficulties, gait disturbance, intractable coughing, or sneezing. Typically, the symptoms do not follow an anatomical nerve distribution nor do the actions fit the symptoms such as a child with a complaint of blindness who does not bump into things. When multiple symptoms are present, it is more suggestive of a somatization disorder.

Although “la belle indifference” is classically associated with conversion symptoms and may be a useful diagnostic sign, it is not a common feature, and the majority of patients with conversion symptoms are in fact upset by them [25].

In a review of over 100 cases of conversion reactions in children, Maloney found that a majority of children came from homes where depression and conflict were present. Almost universally the onset of symptoms was associated with familial stress. Three quarters of the families had difficulty with emotional expression and communication [26].

A Dutch study found a history of physical/sexual abuse in patients with conversion disorders more often than in matched comparison patients [27].

In a Swedish study, children with motor conversion disorders were compared to age- and sex-matched patients with motor symptoms due to a neurological disorder. They found depression, the presence of a personality disorder, and also poor schooling to be significantly associated with motor conversion disorder. Low levels of affection and warmth during childhood along with perceived parental rejection was higher in the group with conversion symptoms. In contrast to other studies, they did not find the history of childhood physical or sexual abuse to be more associated with conversion disorders [28].

Maisami and Freeman demonstrated success in treating children with conversion reactions when child psychiatry and pediatric neurology worked together in the evaluation and treatment. The underlying stress was identified and the treatment emphasized health rather than disease [29]. In a study by Pehlivanturk and Unal, 85 % of children with conversion disorders were symptom free at a 4-year follow-up. A favorable prognosis is associated with early diagnosis and good premorbid adjustment [30]. Crimlisk found that patients who presented with sensory symptoms tended to have better outcomes that those who presented with weakness [31].

The diagnosis of conversion disorder can be a clinical challenge. Making the diagnosis early in the course of the presentation can reduce the child and family's anxiety and reduce the need for costly and unnecessary tests. A thorough physical examination and a careful psychiatric history are essential to screen for comorbid psychiatric illness. The history should include the onset and nature of symptoms and the presence of stressors. Establishing rapport while eliciting the history is very important. The therapeutic relationship between the clinician and the child and family will help them understand and better accept the diagnosis. It is not uncommon for children to be unable to verbally express the psychological factors that are stimulating their symptoms. Their body is expressing it for them [32].

With the advances in neuroimaging techniques, it has been possible to study the neural basis of conversion disorders. Using PET scans, two researchers showed a change in cerebral blood flow in patients with the conversion symptom of hemiparesis: increased cerebral blood flow in the right anterior cingulate and right orbitofrontal cortex; and deactivation of the left dorsolateral prefrontal cortex [33]. Another study using SPECT scanning showed reduced blood flow in the thalamus and basal ganglia contralateral to the deficit, which resolved when the symptoms ceased [34]. This suggests an emotional moderation of motor processes in the striato–thalamo–cortical circuits. It is postulated that emotional stressors inhibit these pathways, which impairs motor readiness and the quality of voluntary movements. Thus, reducing the child's subjective distress may be the most effective treatment for conversion disorders.

Tics and Tourette

Tics are readily observable involuntary sudden, rapid, repetitive, or nonrhythmic stereotypic movements or vocalizations. This neuropsychiatric disorder may also have a variety of concomitant psychopathologies including obsessive compulsive disorder (OCD), attention deficit/hyperactivity disorder (ADHD), learning difficulties, and sleeping abnormalities [35]. There are no laboratory tests for tics and diagnosis is based solely on the history and clinical examination. Tics may be simple, complex, transient, or chronic. Although it is essential for researchers to separate chronic tic disorder from Tourette, which includes both motor and phonic tics, in practice, it has little relevance for outcome or treatment.

A neurological basis for tics with pathophysiologic involvement of several different neurotransmitters has been described. Further, genetic abnormalities that predispose to Tourette Syndrome have been identified [36, 37]. However, it is well recognized that stress and anxiety may exacerbate tic symptoms. It can be challenging to distinguish between tics and behavioral

symptoms [38]. Depending on the study, the frequency of aggression and explosive outbursts of patients with Tourette has been reported to vary from 26 to 75 % [39]. In a study of school aged children in the UK, tics occurred in 65 % of the students with emotional and behavioral difficulties, 24 % of the students with learning difficulties, and in none of the normal children [40]. In a study by Mason, teachers rated children with tics as having more emotional and conduct disorders [41]. The anger dysregulation and outbursts of physical or verbal violence in about 25 % of the clinically referred youth with Tourette may result from disordered impulse control or anxiety disorder. Children with tic disorders have a chronic, socially disabling, and stigmatizing disease. It is not uncommon that they are bullied, which can result in the development of anxiety and depression [42–44].

Jankovic in his description of the phenomenology of tics describes them as both semivoluntary and involuntary and both suppressible and suggestible. Tics do not happen by choice. Yet, with psychological effort, they can be partially controlled or they may be triggered by suggestion. Tics increase under stress and will decrease with distraction and concentration [45, 46]. Children describe the premonitory urge that precede tics and a capacity for brief periods to suppress them [47, 48].

For the clinician, making the diagnosis of a tic disorder includes observation; a review of the developmental, medical, and family history; onset, description and course of tics; and any co-occurring conditions. Identifying Tourette or a tic disorder is a clinical diagnosis based on the enduring presence of motor tic and in the case of Tourette an additional vocal tic. An essential step toward appropriate and effective treatment is determining the degree to which the symptoms are exacerbated by stress, anxiety, or depression. Impaired adaptive functioning may be related to the tic disorder or to the presence of comorbidities such as ADHD, OCD, learning disabilities (LD), and other behavioral difficulties. It is important to ascertain what psychosocial stressors exist for the child and explore the impact of the symptoms on family members, educational success, and peer relationships. Medical management without psychological support is doomed to failure.

How to Make the Diagnosis of a Functional Neurological Disorder

Rickert and Jay developed an interview strategy to approach the evaluation of a child with symptoms that may be consistent with a psychosomatic disorder. Their approach can be remembered by the acronym SAFE (severity, affect, family, and environment) [49].

Severity: When assessing the severity of the symptoms, both the child's description and the manner in which they are communicated are important. A detailed description with a lot of imagery may indicate that the child's symptoms are a coping strategy for emotional distress and worry in the child's life. For the child, having a headache may provide an acceptable excuse for avoiding stressors. Determining the acute, recurrent, or chronic nature of the symptoms is also important.

A functional etiology for symptoms is more likely when there are multiple complaints inconsistent with pathophysiologic principles. When the time and location of symptoms is vague and highly variable or clearly associated with stressors, an organic cause is less likely. Although the emotional distress that can be the stimulus for psychosomatic neurological symptoms can interfere with restorative sleep, the functional symptom itself rarely wakes one from sleep. When there is an underlying organic pathology for the child's symptoms, specific measures such as anticonvulsants for seizures or analgesics for headaches may bring relief. When the symptoms are functional, they are rarely relieved by conventional measures other than by rest and time. Frequently, the child or parent may report "nothing works."

Affect: The next step is to assess how the child and parent have adapted to the child's symptoms. The child with functional neurological symptoms may seem nonchalant and unconcerned that his disabling illness has resulted in an altered lifestyle, school absences, and even bed rest. The patient's flattened affect may represent depression.

It is also important for the evaluating clinician to recognize his or her own affect or gut reaction

to the child, family, and the presenting symptoms. Past experience and a gut feeling may enhance or distort clinical judgment. It is possible and not-uncommon that a child can present with both functional neurological symptoms and have underlying disease.

Family: The expression of a functional neurological symptom may be the expression of a larger family problem. It is easy to recognize a dysfunctional family when immature parents, psychosocial chaos, drugs, alcohol, mental illness, criminality, abuse, and contentious parents are involved. Sometimes, the stress experienced by the child within the family may be more covert. The parent may be over involved in the life of their child, e.g., by pushing the child to overachieve academically, musically, or athletically. Conversely, the parent may be physically or emotionally unavailable to the child and the somatic complaint may be a way for the child to garner attention.

A parent may also model functional symptoms. In that family constellation, having a physical illness may be more acceptable than displaying emotional or behavioral symptoms. The parent may be unwilling to accept the possibility that the symptom may be due to family stressors.

Another significant factor is the role of the family in the secondary gain of the symptom for the family. Perhaps having a "sick" child allows the parent to remain at home, provides distraction from marital issues, or is related to a financial or legal gain. An absent parent may become more involved when their child has neurological symptoms.

How the family responds to the child's symptoms is very important for the clinician to assess. When the family makes too much fuss over the somatic disorder or views a situation as considerably worse than it actually is, the symptom may escalate. This can result in giving the child's neurological symptom too much power within the family. The child may become absorbed in the symptom, making it part of his or her persona. When the family is overly concerned and focused on the somatic complaint, the child may fear that whatever is wrong is life threatening. Conversely, the child's symptoms also may exacerbate and persist when the family ignores the complaints or tells the child that it is being faked.

Environment: Functional neurological symptoms also may be triggered by stressors in the child's environment outside of the family. Stressors at school, with peers, and in the community need to be assessed. When the onset of symptoms happens before or at school, but does not interfere with socializing with peers, school avoidance may contribute to functional symptoms. When symptoms result in not being involved with peers, depression, peer ridicule, and teasing may be significant factors. The relationship between community-related events and the illness should also be explored. Some children may dislike taking music lessons, recreational activities, or athletic competition and their functional symptom is an acceptable means of avoidance. Even musically or athletically gifted children may develop performance-related psychosomatic complaints. Psychogenic neurological problems may also arise following natural disasters or human tragedies (see Chap. 17).

When the complex interaction between the child's emotional and physical state are imbalanced and psychogenic neurological symptoms result, a multidisciplinary team approach to diagnosis and treatment is the most effective. Unnecessary tests, medications, and ineffective treatments can be avoided. The initiating stressors and perpetuating factors can be explored and an effective treatment plan that recognizes the functional nature of the symptoms can be developed [29].

Treatment

The prognosis for children with functional neurological symptoms that are diagnosed promptly generally is favorable. When the symptoms are misdiagnosed, the condition can become chronic, effective treatment is delayed, and the underlying stress is ignored. In such a setting, the prognosis is far less promising. For the clinician, the child's treatment begins with nonjudgmental acceptance of the child regardless of the nature of his or her functional neurological symptoms.

For many parents and some children, it is easier to accept a physical explanation for neurological symptoms. This is especially true when a parent is the source of much of the child's stress. There may also be the fear that an ominous physical cause may be overlooked and perhaps more testing should be done. The child and the family need an understandable explanation, reassurance, and support.

The clinician can assist the family in arranging counseling for the child, which can incorporate cognitive and behavioral therapies (see Chap. 19), psychological resiliency, and training, if needed, and skills to help them in social situations. Family counseling may also be beneficial. Parenting classes and discipline training can help the parent recognize that the underlying purpose of discipline is to instill a sense of self-control and responsibility for one's behavior.

It is important to involve the schools in the treatment plan. Teachers, coaches, and school nurses must demonstrate acceptance and appropriate management of the child's symptoms. There needs to be open and ongoing communication between school, home, and the multidisciplinary clinician team. It is necessary to explore and mitigate the amount of stress the child experiences at school, e.g., safety concerns, bullies, teasing, learning disabilities, and too much pressure to perform.

Rather than focusing on the functional disorder, the clinician, family, and school should look at the whole child, especially his or her talents, skills, and interests. For the child, recognizing his or her own strengths can help develop self-esteem that may ultimately lead to self-mastery over symptoms.

Given the modest response of most pharmacotherapies for functional neurologic disorders, the considerable side-effect profile, and the fact that the most significant and bothersome symptoms are usually triggered by stress, it is important that the child with functional symptoms or symptoms with an organic basis that are exacerbated by stress be taught relaxation skills such as with breathing, hypnosis (Chap. 21) and self-regulation strategies such as biofeedback (Chap. 20).

Case Studies

Case 1: Tic Disorder

SC was a 17-year-old with Tourette Syndrome that began at age 12. He also experienced anxiety and attention deficit hyperactivity disorder. He had significant motor and vocal tics which exacerbated when he began his high school years at a boarding school. His polypharmacy included medications for anxiety, tics, and attention-deficit disorder. As he settled into school and made new friends, his tics improved. Now in his senior year, he was accepted into his first choice college. His tic frequency has escalated significantly.

Questions

1. What is the most likely cause of the exacerbation in his symptoms of Tourette?
   (a) Noncompliance with prescription medications
   (b) Change in medication
   (c) Anxiety about transition to college
   (d) Self-medicating with drugs and alcohol
   (e) ADHD medication
2. In addition to ADHD and anxiety, all of the following are often comorbidities of tic disorders EXCEPT:
   (a) Learning disability
   (b) Obsessive–compulsive disorder
   (c) Impulsivity
   (d) Dissociative identity disorder
   (e) Emotional lability
3. What is the most appropriate therapy for this patient?
   (a) Hypnosis
   (b) Increase his medication
   (c) Cognitive behavioral therapy (CBT)
   (d) (a) and (c)
   (e) (a), (b), and (c)

Answers

1. (c): Anxiety about transition to college Over the course of his 4 years at boarding school, SC had developed a great group of friends who accepted his tics and even explained his

behavior to curious new students. SC was very anxious about the upcoming transition to college and was worried that he would have to explain his Tourette to everyone. There were no new medications or change to his pharmacotherapy. There is no evidence (group data) that motor tics or vocal tics change in frequency or severity during maintenance therapy. SC was not drinking or smoking.

2. (d): Learning disability, obsessive compulsive disorder, impulsivity, and emotional lability are all comorbidities of tic disorders.
3. (d): Hypnosis can provide SC with self-regulatory strategies to help him better manage his tics. CBT can be an effective therapy for symptom-based diagnoses integrating behavior therapy with cognitive therapy.

Case 2: Nonepileptic Seizures

BF suffered a traumatic brain injury when he was physically abused by his mother's boyfriend at age 6. He subsequently developed a partial seizure disorder, which was well controlled on an AED. When his mother was incarcerated on drug charges, his grandmother became his legal guardian. At age 12, he had his first seizure in 4 years. His AED was changed. Two months later, he had what appeared to be a tonic–clonic grand mal seizure. His mother was scheduled to be released from prison that week. His AED was increased, but the tonic–clonic seizures continued sporadically.

Questions

1. What might suggest that the new seizure was an NES?
   (a) Psychosocial stress
   (b) Change in seizure type
   (c) PTSD
   (d) No improvement on AEDs
   (e) All of the above
2. Characteristics of NES include all of the following EXCEPT:
   (a) Rise in prolactin level
   (b) Gradual onset
   (c) No clear loss of consciousness
   (d) No incontinence
   (e) No postictal confusion
3. Which of the following will provide the most definitive evidence that this new seizure is nonepileptic?
   (a) Video EEG
   (b) CT scan
   (c) MRI
   (d) EEG
   (e) Spinal tap

Answers

1. (e): All are historical clues that might lead the clinician to suspect NES
2. (a): Prolactin levels rise after epileptic seizures. The remaining choices are all clinical clues that suggest NES
3. (a): Video EEG is considered the gold standard for diagnosing NES. In the absence of clinical seizure activity, a normal EEG does not exclude the possibility of ES or NES.

Case 3: Headaches

JJ was just 9 years old when her mother died of cancer. Her father was still serving at least 10 more years of a prison sentence, so she and her two younger brothers moved in with her paternal grandparents in another state. The grandparents were poor, older, not in good health and overwhelmed by the addition of three children to their home. JJ had mild mental retardation, microcephaly, obesity, enuresis, and encopresis. One of the brothers had attention deficit hyperactivity disorder and both were on the autism spectrum.

During JJ's teenage years, her grandparents separated and her grandmother's health deteriorated. In addition to the expectation that she maintain her attendance at school, JJ was expected to do the cooking, cleaning, laundry, and care for her grandmother. Her brothers were of little help and she argued continually with one of her brothers. JJ had few friends, school was a struggle, and she had little time for pleasurable pursuits. JJ became depressed and experienced daily headaches that with a flat affect she described as a 10 on the 0–10 pain scale with zero being no pain and 10 the worst pain in the world that she can imagine. Her headaches were unresponsive to OTC analgesics. With the amount of

psychosocial stress in her life, a psychogenic headache was the obvious diagnosis.

Questions

1. Although psychogenic headaches is the likeliest primary diagnosis, what else needs to be considered?
   (a) Sinusitis
   (b) Increased intracranial pressure
   (c) TMJ dysfunction
   (d) Migraine Headache
   (e) All of the above
2. What possible reason could explain the lack of response to OTC analgesics?
   (a) She had a bacterial sinusitis
   (b) Ongoing psychosocial stress
   (c) Depression
   (d) Rebound from overuse of acetaminophen
   (e) All of the above
3. What is an appropriate next step in this patient's management?
   (a) Involve social services
   (b) Teach JJ self-regulation strategies
   (c) Mental health counseling
   (d) Improve support systems
   (e) All of the above

Answers

1. (e): All need to be considered in the differential diagnosis. Surprisingly, a CT revealed extensive sinusitis. Her headaches improved somewhat after antibiotic therapy, but did not resolve.
2. (e): All are possible explanations for her lack of response to OTC analgesics.
3. (e): Multiple agencies and services are often necessary when the child's psychosocial issues are significant.

Conclusions

When the complex interaction between the child's emotional and physical state is imbalanced, psychogenic neurological symptoms may result. When stressed, children may develop functional symptoms or the expression of a true organic condition may change or exacerbate. The frequency of headaches for the child with migraines may increase. The child with epileptic seizures may additionally develop nonepileptic seizures. The child may convert their psychologic stress into sensory or motor dysfunction. Tics can certainly be exacerbated in response to stress. Medications used to control seizures and tic disorders have significant side effects. Medical and pharmacologic management without an accurate diagnosis and understanding of the functional basis of the symptoms likely will have little benefit and may actually cause harm.

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Chest Pain, Syncope, and Palpitations in the Pediatric Patient

3

Frank Coshey Smith

Abstract

Chest pain, syncope, and palpitations are the most common chief complaints in children that might be cardiac in origin. Although all three are often associated with significant heart disease in adults, they usually represent benign conditions in children, do not require aggressive treatment, and may be considered to some degree "functional." Nevertheless, parents of children with any of these complaints are often anxious that their child may be the exceptional case with life-threatening (usually cardiac) disease. The purpose of this chapter is to provide the primary care practitioner with a practical office approach to the child with chest pain, syncope, or palpitations of a semiacute or chronic nature. For each complaint the most common noncardiac, as well as the rarer cardiac causes will be reviewed. A discussion follows that emphasizes important aspects of the history and physical examination that will help to differentiate cardiac from less serious, noncardiac causes. The main objective is to help the practitioner to evaluate each complaint effectively, reassure patients and families appropriately, and avoid unnecessary testing and referrals to an emergency department or subspecialists.

Keywords

Anxiety • Chest pain • Children • Palpitations • Syncope

The most common chief complaints of a child that might be cardiac in origin are chest pain, syncope, and palpitations. Although all three are often associated with significant heart disease in adults, they usually represent benign conditions in children, do not require aggressive treatment, and may be considered to some degree "functional."

Nevertheless, parents of a child with any of these complaints are often anxious that their child may be the exceptional case with life-threatening (usually cardiac) disease. When these problems present acutely and with intensity they are often evaluated in an emergency department (ED, Chap. 15). When they present with less intensity or recur over

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a longer period of time, they are usually evaluated in a primary care provider's office.

The purpose of this chapter is to provide the practitioner with a practical office approach to the child with chest pain, syncope, or palpitations of a semiacute or chronic nature. For each complaint, the most common noncardiac as well as the rarer cardiac causes will be reviewed. Important aspects of the history and physical examination will be discussed that help the practitioner to differentiate cardiac from less serious noncardiac causes. Each section concludes with advice about how to inform and reassure the patient and family when the presenting problem is unlikely to be due to a cardiac or other significant cause and will not require management beyond simple, nonspecific or preventive measures.

The main objective is to help the generalist to evaluate each complaint effectively, reassure patients and families appropriately, and avoid unnecessary testing and referrals to subspecialists. The information may likewise prove useful for pediatric cardiologists and other subspecialists who are referred children with these complaints and are expected not only to exclude significant medical causes for the complaints but also to reassure the patient and family definitively that the complaint does not require specific therapy and is, in effect, functional.

Chest pain, syncope, or palpitations in a child *with previously diagnosed congenital or acquired heart disease* presents a special situation. Although the child's primary care provider may apply the following information to an initial evaluation of the child's complaint, a pediatric cardiologist will likely be consulted and further management may require testing and treatment that would otherwise not be indicated for the child without a history of heart disease.

Chest Pain

Causes

Of the three chief complaints discussed in this chapter, chest pain is probably the most common and occurs at some time in nearly 10 % of all school age children. Nevertheless, its cause is usually noncardiac and often benign [1, 2]. Chest pain in a child may be of musculoskeletal, idiopathic/psychological, pulmonary, gastrointestinal, or cardiac origin. Comprehensive lists of all potential causes of chest pain have been described previously in the literature and particularly in the emergency department setting [3, 4]. In a recent review of 3,700 consecutive cases of chest pain in children, the most common causes in descending order of frequency were idiopathic, musculoskeletal, pulmonary, gastroenterological, cardiac, anxiety, and drug related [1].

Idiopathic chest pain in children is often attributed to musculoskeletal causes or it may be considered psychogenic. Musculoskeletal conditions are the most common identifiable causes. These often involve strain from previous trauma, repetitive exercise, sports participation, or perhaps simply from somatic growth. Other causes include floating rib syndrome or costochondritis. Musculoskeletal pain may occur at any place in the chest. It is presumably due to inflammation and can occur at anytime, during rest or activity. It is usually a sharp or crampy pain and it may be worsened with further strain on the affected area by exercise, deep breathing, or applied external pressure. Since the pain is not vascular, it does not usually radiate to the arm, shoulder, or wrist. The pain may only last a few seconds at a time. A history regarding gym and sports participation, repetitive exercises, heavy lifting or carrying, or wearing a tight backpack should be sought. Clinical findings include evidence of previous chest trauma, tenderness to palpation over the area of pain, mobile lower ribs, and reproduction of the pain with various arm and chest movements by the patient. The pain may be reduced by analgesics and anti-inflammatory medications.

Pulmonary conditions are the second most common cause of chest pain in children. Acute causes include asthma, pneumothorax or pneumomediastinum, pneumonia, pneumonitis, pleuritis, and pleurodynia, pulmonary embolism, and chest tumors. Many of these causes will be identified by additional history of other pulmonary symptoms, abnormal findings on clinical examination of the lungs, or abnormal chest radiograph

or other ancillary tests. Of these causes, exercise induced asthma may be the most difficult to differentiate from a cardiac cause since it occurs specifically with exercise. In these patients, there is usually a history of asthma and sometimes the previous use of a nebulizer to control the pain. Pneumothorax and pneumomediastinum may occur in association with acute asthma. The associated pain is usually excruciating, worsened with deep breathing, and associated with typical findings on chest examination. Pneumonia and pneumonitis are associated with fever, cough, and constitutional symptoms. Clinical examination may reveal rales, diminished breath sounds, or wheezes. Pleuritis is relatively rare but may complicate viral upper and lower respiratory infections. Pain attributable to pleuritis is particularly sharp, is located over one or both sides of the chest or back, and usually worsens with inspiration. It usually occurs at rest as well as with exercise. Pleurodynia is a condition that is not well described or understood. It is associated with severe, sharp pain in the lower midsternum that frequently causes the child to "double over" and is more likely to lead to an emergency department visit than other causes of chest pain. Pleurodynia may be associated with pallor and transient respiratory distress. It is relieved with rest and then subsides for hours or days. Episodes may occur over several weeks. Treatment of pleurodynia is reassurance and simple analgesia with acetaminophen and ibuprofen. Pulmonary embolism is rare in children but may occur in teenagers with certain risk factors (e.g., severe obesity, prolonged immobilization after an injury or operation, or hypercoagulable states). The chest pain is usually associated with dyspnea and patients may also be hypoxemic or have abnormal findings on chest examination or chest radiography. Chest tumors are a very rare cause of chest pain. Pain attributable to tumors usually is more persistent and unrelated to exercise.

Gastroesophageal reflux is associated with an epigastric and sometimes substernal pain that usually has a burning sensation. The pain may radiate upward toward the throat. It may be associated with certain foods and other gastrointestinal symptoms. Although it is commonly referred to as "heartburn," it is usually easy to differentiate from other causes of chest pain and may respond quickly to antacids or more advanced treatment of gastroesophageal reflux disease.

Cardiac causes of chest pain are the least frequent in children. These causes can be divided into those related to reduced coronary artery perfusion, inflammation of the heart, or miscellaneous causes. Reduced coronary artery perfusion may be associated with left ventricular outflow obstruction or coronary artery anomalies. The most common causes of left ventricular outflow obstruction are congenital valvar aortic stenosis and subvalvar aortic stenosis secondary to hypertrophic cardiomyopathy. Hypertrophic cardiomyopathy is a disease associated with progressive left ventricular hypertrophy and asymmetric ventricular septal hypertrophy that usually becomes apparent during the second decade of life [5]. It is familiar with autosomal dominant inheritance. It should be considered in any patient with a family history of unexpected sudden death or any cardiomyopathy. It may present with a murmur of subaortic stenosis, chest pain, syncope, exercise intolerance, or arrhythmias. It is the most common cause of sudden unexplained cardiac death in the adolescent and young athlete. For this reason, this is certainly a diagnosis that all clinicians must consider in the evaluation of any adolescent with chest pain, as well as syncope or palpitations. In this case, the obstruction occurs between the hypertrophied ventricular septum and anterior mitral valve leaflet. The murmur of obstruction is a harsh systolic ejection murmur heard at the lower left sternal border. Since the obstruction increases with transient decreases in ventricular volume, the murmur characteristically becomes louder when the patient stands or performs the Valsalva maneuver. The diagnosis may be suggested by left ventricular hypertrophy or ST-T waves changes on an electrocardiogram (ECG) and it is confirmed by echocardiography.

Coronary artery anomalies include congenital abnormalities of the exact location of the coronary artery origins from the aorta [6]. These are associated with ischemia and cardiac symptoms during exertion. An equally rare cause is coronary artery thrombosis in a child with a

history of Kawasaki Syndrome and large coronary artery aneurysms [7]. In cases of limited coronary artery perfusion, the chest pain is usually, but not always, similar to angina pectoris of the adult with coronary artery disease. The pain is midsternal and it is usually described as a pressure or crushing sensation, not sharp. The pain may radiate to the left shoulder or left arm. The pain tends to persist for more than a few seconds. It is typically only exertional and persists until the patient rests. It is not affected by deep breathing or applied external pressure, or particular movements of the arms or chest. It is usually not relieved with analgesics. In addition, the child may have pallor, diaphoresis, shortness of breath, or abdominal pain [7].

Inflammation of the heart (pericarditis or myopericarditis) causes a rare and distinctive type of chest pain that is usually acute and more likely to present as an emergency. It is usually viral or rarely bacterial in origin, or it may be a manifestation of more widespread chronic inflammatory disease. Inflammation of the visceral and parietal pericardium leads to severe pain that is usually sharp, mid sternal, or left parasternal. The pain varies not only with deep breathing but also with changes in body position. A pericardial friction rub may be heard at the lower left sternal border or apex. This is a superficial, "scratching" sound that usually has components in systole and diastole. Since the cause is usually infectious, the child often has a fever and other constitutional symptoms. In cases associated with a large pericardial effusion, muffled heart sounds and signs of pericardial tamponade are observed. Myocarditis often presents with nonspecific symptoms and signs in younger children, but it may present with chest pain in older children. The pain is usually associated with respiratory and gastrointestinal symptoms and signs and also presents more often to the emergency department. It is usually associated with ECG abnormalities and elevation of cardiac enzymes [8].

Miscellaneous cardiac causes of chest pain include pulmonary hypertension, aortic dissection, arrhythmias, and premature extrasystoles. Although dyspnea on exertion is the most common presenting complaint due to pulmonary hypertension in young children, syncope and chest pain are also common complaints [9]. The pain and associated symptoms are usually related to exertion. The pain is substernal in location and may present either as an emergency or subacutely. Examination may reveal a narrowly split second heart sound, a prominent second (pulmonary closure) component to the second heart sound, or a completely single second heart sound. Rarely, a high-pitched diastolic murmur of pulmonary incompetence may be heard at the upper left sternal border.

Chest pain caused by aortic dissection is usually intense and leads to emergency department evaluation. Rarely, the dissection may be subacute with less intense, chronic symptoms. The diagnosis is usually associated with Marfan syndrome or previously diagnosed aortic valve disease, or it can be a familial disorder in the absence of any obvious connective tissue disorder [10].

Premature ventricular contractions (PVCs) may present with palpitations or detection of an arrhythmia during a physical examination, but they may also be associated with chest pain [11]. The pain is often described as twinge-like and sharp, lasting a few seconds, and may be associated with a subjective awareness of a single strong heartbeat. These are not usually related to exercise. In fact, benign PVCs occur more commonly at rest with slower sinus rates and are suppressed at higher sinus rates (e.g., during exercise). PVCs should be confirmed by electrocardiography. Consultation with a pediatric cardiologist, exercise testing, Holter or event recordings may be necessary in problematic cases [11].

In the recent review of 3,700 cases of children cited earlier with chest pain, a significant cardiac cause was demonstrated in only 0.8 % of cases. The causes in this review included aberrant origins of a coronary artery, pericarditis, myocarditis, and cardiomyopathy [1]. In another recent study of 4,436 children with chest pain who presented acutely to a pediatric emergency department, a cardiac cause of the pain was confirmed in only 0.6 % of the patients [12].

Idiopathic chest pain in children may be related to minor musculoskeletal strain that is not detectable by history or physical examination alone.

Some of these children may have a psychological basis for the symptom. In one study, a small cohort of children and teenagers with noncardiac chest pain underwent psychological evaluation. A current DSM-IV Axis I panic or anxiety disorder was detected in 59 % of the patients [13]. In a later study, patients referred to a pediatric cardiologist with chest pain were significantly more likely to have a panic or anxiety disorder than those who were referred for evaluation of a heart murmur [14]. These patients would likely benefit the most from psychological counseling and behavior modification [15].

History

Although there are few studies that have identified specific details of the history that accurately differentiate cardiac from noncardiac chest pain, the presence of pain with significant exertion or pain that awakens the child during sleep have been associated more frequently with cardiac etiologies [1, 4]. Nevertheless, taking a comprehensive history about chest pain serves several purposes. First, it may shed light on the correct cause of the pain. In addition, and equally importantly, it inspires confidence since it communicates to the family from the beginning of the evaluation that the problem is being treated seriously and carefully.

A proper history depends not only upon the physician's ability to know what to ask, but *how to ask it*. Questions are best asked in an unhurried, organized fashion. Consistency in questioning should be maintained from visit to visit and from patient to patient in order to insure thoroughness.

Onset: When did the pain first occur? In most cases of nonemergent chest pain, the first occasion is one of several episodes that occurred often weeks or months before the evaluation.

Location: Ask every patient to point with one finger to the place where the pain hurt the most. Cardiac pain tends to be midsternal, or less likely left parasternal. The majority of noncardiac chest pain is left parasternal or more lateral under the left nipple, or elsewhere. In general, the farther from the sternum the pain is felt, the more likely its cause is noncardiac.

Quality of the pain: Although open-ended questions are optimal, many children will shrug their shoulders when asked "What does the pain feel like?" In that case, provide possible choices for an answer while making every effort to avoid "leading the witness." For example, asking "Does it feel as if someone is sitting on your chest pressing it?" while nodding vigorously may encourage the child to answer affirmatively when this really is not the case. When the child cannot describe the pain voluntarily, present possible choices in a dispassionate manner: "Does it feel as if someone is sitting on your chest, or does it feel sharp, or does it feel like a burn…?" The majority of children and their families will respond "sharp" once this option is presented because musculoskeletal and idiopathic chest pain is most common and it is usually sharp. Cardiac pain with the exception of pericarditis is not sharp and is more of a pressure sensation.

Effect of exertion: Again, provide optional responses in a dispassionate way. Ask "Does the pain hurt more at any particular time? Does it seem to hurt more when you're playing really hard, or more when you're sitting around resting, or can it happen at any time?" Cardiac pain is typically associated with exertion. Noncardiac causes except exercise induced asthma are not.

Effect of body position: Pericardial pain is particularly affected by body position or changes in position. Other cardiac causes and all noncardiac causes usually are not.

Radiation: Asking "Does the pain radiate anywhere?" is unlikely to provide accurate information since most families are not familiar with this medical term. Asking the patient whether the pain "shoots," "jumps," "moves," or "spreads" to a different place, again while providing choices for destinations like the arms, legs, or head is more likely to provide useful information.

Cardiac pain may radiate to the left shoulder or arm. Pericardial pain may radiate to either shoulder. Noncardiac pain does not radiate unless the child has several points of musculoskeletal sprain.

Effect of breathing: Avoid leading questions such as "Does it hurt more when you take a deep breath?" and ask instead "Does deep breathing do anything to the pain—make it worse or better?" This provides more reliable information. If the pain is altered in any way by deep breathing, it is more likely to be due to musculoskeletal or pleuritic rather than cardiac disease.

External pressure: Ask "Does the pain change if you touch or massage the place where it hurts?" If the patient reports that the pain changes significantly with applied pressure, this is clearly musculoskeletal and not cardiac in origin.

Associated symptoms and signs: At this point, the reports of the parents may be helpful, if they have not already contributed to the history above. Observed dizziness, breathlessness, pale color, perspiration, or any other signs of distress, may indicate a significant cardiac or pulmonary cause.

Intensity: A question often asked as part of the emergency department evaluations of children with chest pain is, "On a scale of 1–10, how bad is your pain?" This approach may be relevant in assessing whether the pain is worsening or improving, but it is the author's impression that it is of no use in the office diagnosis of chest pain since it cannot be standardized from patient to patient. Furthermore, whatever the number assigned to it, the pain was intense enough to warrant an evaluation by a physician. If the child reports pain during the office evaluation, the vital signs, heart, and lung examination should be performed immediately with attention paid to the heart rate and rhythm, cardiac and lung auscultation. An attempt by the examiner to reproduce the pain by applying external pressure to the area or having the patient take deep breaths may be helpful to determine its origin. If heart rate is not elevated and the facial appearance and behavior of the child do not indicate significant pain or stress, this may also help to confirm a benign cause of the pain.

As noted above, a positive past medical history of significant cardiac disease should lead to early consultation with the pediatric cardiologist. History of chronic inflammatory disease could be associated with pericarditis/myocarditis [8]. Past history of chronic pulmonary disease would raise the possibility of a pulmonary cause or perhaps pulmonary hypertension. History of scoliosis, skeletal deformities, or chronic muscular disease could indicate a musculoskeletal cause or perhaps a connective tissue disorder that is associated with cardiac disease.

Family history of any "heart muscle disease" (i.e., cardiomyopathy), unexpected sudden death or heart attacks at an early age, or the known diagnosis of hypertrophic cardiomyopathy should be excluded. Other familial causes of sudden death, particularly congenital long QT syndrome, might be associated with arrhythmias that produce chest pain, as well.

Physical Examination

Physical examination of the child with chest pain should be complete, but with specific attention paid to examination of the chest. This should include assessment and palpation of the rib cage and sternum for any deformities, bruises, mobile ribs, areas of swelling, or reported tenderness. Pulmonary assessment should include assessment of air entry in all lung fields and the presence of any adventitial sounds. Any abnormal findings should lead to further assessment of a pulmonary cause. The cardiac examination should include palpation of the precordium for any hyperactivity, thrills, or displacement of the apex beat laterally. The first and second heart sounds should be identified and physiologic splitting (separation of the aortic and pulmonic closure sounds during inhalation) should be assessed. The presence of additional heart sounds should be assessed, noting any murmur, click, rub, or gallop and whether any murmur changes in intensity when the child is sitting, supine, standing, or performing the Valsalva maneuver.

Hypertrophic cardiomyopathy is often associated with dynamic obstruction of left ventricular outflow during systole that is louder with standing or during the Valsalva maneuver. Rarely, only a gallop rhythm may be heard with this diagnosis. Other causes of aortic obstruction (subaortic stenosis, valvar aortic stenosis, etc.) are associated with loud systolic murmurs that have usually led to a cardiac diagnosis in early childhood. The patient with pericarditis may be uncomfortable during the examination because the pain varies with changes in body position. A pericardial friction rub, a superficial scratching sound in systole and diastole, may be audible at the lower left sternal border or apex. In cases with significant pericardial effusion and pericardial tamponade, there may be hepatomegaly, jugular venous distension, and an exaggerated reduction in pulse volume during inspiration (pulsus paradoxus).

Additional Tests

A chest radiograph should be considered in any child with chest pain who has definite pulmonary symptoms or abnormal physical signs on lung examination. When a cardiac cause of chest pain is suggested by history or physical examination, an electrocardiogram may help to rule in specific cardiac causes. Some generalists order an ECG in all children with chest pain simply to exclude obvious signs of hypertrophy related to hypertrophic cardiomyopathy, or signs of preexcitation (Wolff–Parkinson–White syndrome), or long QT syndrome.

The decision process regarding which children with chest pain should be referred to a pediatric cardiologist varies among physicians and regions of the country. The incidence of significant heart disease among children with chest pain is very low and the cost per patient to exclude heart disease with imaging tests is relatively high. A Standardized Clinical Assessment and Management Plan (SCAMP) has been implemented at Boston Children's Hospital in an attempt to identify children with chest pain who are more likely to have cardiac disease and limit testing in those children who do not. This includes a thorough past medical and family history of each patient and prioritizes children with exertional chest pain from those without it. A study in which the chest pain SCAMP was implemented reduced the total cost of evaluating children with chest pain by 21 %, without adversely affecting diagnostic accuracy [16].

Discussion with Patient and Family

Cases of chest pain with features of a pulmonary or gastrointestinal origin may be treated by the generalist or referred to the appropriate specialist for evaluation. Children should be referred to a pediatric cardiologist if there is a high suspicion of cardiac disease after performing a thorough history and physical examination, if additional tests such as a chest radiograph or ECG are abnormal, or if the practitioner cannot completely reassure the patient or family that the pain is not cardiac in origin.

In brief, chest pain that is sharp, nonradiating, located lateral to the sternum, of short duration, or is affected by deep breathing or external pressure, but unassociated with exercise or pulmonary symptoms is very unlikely to be cardiac or pulmonary in origin in an otherwise normal child. The most likely cause of noncardiac chest pain is musculoskeletal or idiopathic. This can be considered "functional" in a sense because the pain is commonly attributed to everyday movements, stresses, and strain of the functioning body.

When the child's chest pain is deemed most likely to be musculoskeletal, the physician can reassure the child and family that the pain is not serious and is most likely related to everyday stress and strain of the muscles, joints, bones, and nerves within the chest. Simple analgesics and local applications of heat may be helpful at times of pain. Temporary limitation of exercise or certain activities may be helpful in cases that are obviously related to muscular strain [3]. If the generalist still detects parental or patient concern specifically about cardiac disease, then an ECG and/or chest radiography may be appropriate and provide further reassurance. In addition, scheduling a follow-up appointment in 2–3 months might provide further reassurance to both the family and the physician.

Table 3.1 Summary of advice for primary care providers: counseling children and families about symptoms of chest pain, syncope, and palpitations that are deemed to be "functional"

Complete a consistently thorough, well-documented history and physical examination. This not only ensures an accurate diagnosis but also inspires the family's confidence that the complaint is being treated seriously and carefully
A careful, caring, and unhurried approach to the problem will often lead to the family's acknowledgment of this and their satisfaction with your diagnosis
Perform well-selected additional tests as needed to exclude significant medical causes of the symptom
Reassure the child and parents that they are not alone. All three symptoms are very common in growing children and adolescents
Acknowledge that the symptom may indeed be the first sign of a serious heart problem in adults, but that clinical experience and many published studies have shown that it is almost never due to a significant heart problem in children and teenagers
Parents and clinicians alike may be understandably concerned that a cardiac diagnosis with life-threatening implications will be missed. It may be helpful for some families to be informed that sudden unexpected cardiac death in athletes is fortunately very rare (estimated 1/200,000 high school athletes per year). Explain that your complete, well-documented cardiac history and physical examination have lowered their child's risk of heart disease even further [17]
Some families are reassured to know that the child's history and physical exam have given you important information ("clues") that the pain is not due to a serious heart or lung problem (e.g., chest pain over the lateral aspect of the chest, dizziness only with abrupt standing). Avoid identifying the information specifically in case the power of suggestion might lead to alteration of the child's reported symptoms in the future
If the parents appear to be reassured from your evaluation, then they will usually be able to reassure their child. Enlist and encourage the parents to be prepared to provide further reassurance to their child if the symptom were to recur
Some parents may inquire insightfully whether their child's symptom is anxiety related. Indeed, symptoms may be intensified by the child's mental state, and this should be explored further and may benefit from psychological support [13]. Stress from schoolwork, gym class performance, or relationships with friends and family could lower the child's threshold for symptoms and the ability to cope with them
The primary physician will often be aware of possible stress within the child's family. Stress at school, and particularly in gym class, may be difficult for a child to acknowledge, however. If this is suspected, the author will often say, "How are you doing in gym class? You know, I never liked gym class very much in high school. What do you think about it?" "Gym class" could be substituted with "math class" or a variety of other activities, depending upon the history and potential issues affecting the child. By acknowledging (and perhaps even feigning) one's own dislike or anxiety for a particular class or activity, the physician may give the child more confidence to express his or her own anxieties
Sometimes, the child will share information that not only may be helpful in understanding the child's symptoms but may also lead to identification of particularly stressful circumstances at home or in school that require intervention
If continued anxiety is sensed in the child or parents, ask the family, "Is there something that you are particularly worried that your child may have?" This may provide insight about misconceptions of the parents that can be resolved by additional explanation, simple tests, or consultations
If the symptoms persists in spite of reassurance, simple analgesics, and time, then referral to consultants and/or for psychological evaluation and support is indicated (Chap. 18) [13–15]

How should the physician counsel the child and family about chest pain that is likely to be "functional?"

Table 3.1 presents an approach for counseling regarding "functional" cardiac symptoms. It may be helpful to explain that chest pain is very common in children and that the child is in the "fortunate majority" of children with chest pain that is functional. Chest pain is an understandable phenomenon in a child since it is usually associated with everyday stress and strain on the muscles, cartilage, bones, and nerves that would be expected in any active, growing child. Many parents will interject that the pains are what older relatives have called "growing pains." It is appropriate to acknowledge that musculoskeletal pain can be quite intense for some children, and that it is helpful for the child and parents to know that the pain is nevertheless not harmful. The family and child should be prepared to expect that the pain is likely to occur again since the child continues to grow and remain physically active [18]. Acetaminophen or ibuprofen may be helpful in some children who have recurrences. This information will

reassure the majority of parents who can then help their child to cope with future episodes of pain, avoiding repeat visits to the physician, emergency department, or specialist.

Some parents may respond defensively, "So you're saying it's all in my child's head!" The author usually responds that the pain is real and is definitely in the chest, but not in the heart or lungs where urgent and possibly invasive and painful treatments would be needed to make it better. Dismissing the symptoms completely, as if fabricated, should be avoided.

Since chest pain implies potentially activity-limiting cardiopulmonary disease to many children and their families, it is not surprising that children with chest pain might have underlying anxiety related to sport activities or gym class. Children who either do not like to participate in gym class because they are particularly nonathletic, have issues regarding their body self-image, or are under pressure from their family or coaches to perform in sport events might be particularly sensitive to chest pains or other cardiac symptoms during exercise.

After children with chest pain are evaluated by a pediatric cardiologist and found to have noncardiac chest pain, the majority have a reduction in the frequency and severity of pain in later follow-up and significant heart disease is virtually never detected later [18]. Psychological counseling may be helpful in some cases (Chap. 18). One preliminary study indicated that children with noncardiac chest pain who have an associated psychological diagnosis may have significant relief of the pain after even brief psychological treatment [15].

Case Study 1

A 14-year-old boy presented to the office of his primary care provider with an acute onset of chest pain while playing intramural basketball. The pain was midsternal, nonradiating, and it was described as a pressure sensation. It improved when he sat down on the sidelines. The pain happened to a lesser degree during basketball the week before. It was not associated with meals. It did not change with respirations, body movements, or external pressure to the location of the pain. He had no recent fever, cough, or wheezing. Cardiac examination revealed a grade I-II/VI systolic ejection murmur at the lower left sternal border that was louder when he was standing.

Question

1. What would be the next step in the evaluation of this patient?
   (a) Chest radiograph
   (b) Electrocardiogram and cardiology consultation
   (c) Trial of bronchodilator before exercise
   (d) Trial of antacids
   (e) Reassure and schedule follow-up in 6 months

Answer

1. (b): The patient's pain has many characteristics suggesting a cardiac cause (exertional, midsternal, pressure-sensation, relieved by rest). In addition, the examination reveals a systolic murmur that is louder with standing, which is the typical murmur of hypertrophic cardiomyopathy with obstruction. The pain is not associated with wheezing or meals, making pulmonary or gastrointestinal causes even less likely. The provisional diagnosis is hypertrophic cardiomyopathy with obstruction and further cardiac tests and a cardiology consultation should be arranged.

Case Study 2

A 13-year-old boy returned to the pediatrician's office with recurrent chest pain. His pain began 2 years ago and after 1 year of intermittent pain he was referred to a pediatric cardiologist. The pain was felt over the left side of the chest, did not radiate, was not related to physical exertion, and was sharp in quality. Episodes of pain lasted 5–10 s and it was sometimes difficult for him to breathe because this made the pain even sharper. The pediatric cardiology evaluation included a complete history, physical examination, ECG, and echocardiogram and was normal. The pain subsided, but

has now recurred over the last month, particularly during hard soccer team practices. The characteristics of the pain and physical examination are unchanged. The child was taken out of gym class and sports by the gym teacher until a physician reevaluated him. Since then, he has had no pains. Soccer season has begun and he is one of the better players. His mother was hoping that you would be able to release him to participate in sports.

Questions

1. Which feature of the pain might suggest a new or previously undiagnosed cardiac problem?
   (a) Left-sided
   (b) Nonradiating
   (c) Sharp sensation
   (d) Occurred during exertion
   (e) Worsens with breathing
2. What additional information would be most helpful in the further evaluation of his pain?
   (a) Intercurrent family history of heart disease
   (b) Repeat echocardiogram
   (c) Oral intake before exercise
   (d) Child's attitude about returning to soccer
   (e) Lab tests for inflammatory markers

Answers

1. (d): The other characteristics are all typically associated with musculoskeletal or idiopathic pain rather than cardiac.
2. (d): The child previously underwent a pediatric cardiology evaluation that included ECG and echocardiography. The most common cardiac causes of chest pain would have been detected during that first evaluation. GE reflux related to eating before exercise is unlikely, given the characteristics of the pain. Tests for chronic inflammatory disease in an otherwise healthy patient would likely be normal. When the child was asked about wanting to return to soccer, he was reluctant to answer. When asked whether there were particularly new difficulties or challenges with soccer, his mother then admitted that his father was the soccer coach and that he had been pushing his son to excel more than ever. Although the child was unlikely to be malingering, his chest pain was likely magnified by the stress that he faced during his practices. His mother spoke to his father who then spoke to the pediatrician and it was agreed that the child could return to all activities, but that he would be coached on the team less aggressively.

Syncope

Syncope is the sudden loss of consciousness and body muscle tone due to a transient decrease in cerebral perfusion. The most common type of syncope in the older child and teenager is vasovagal syncope and its course and prognosis are usually bening. It is estimated that at least 15 % of adolescents have had at least one syncopal event. Other causes of syncope are neurological or, most rarely, cardiac [19, 20].

A discussion of the etiology of vasovagal syncope is beyond the scope of this chapter, but the ultimate abnormality is a transient decrease in blood pressure and a decrease (or occasionally an inappropriate increase) in heart rate. This may be precipitated by activation of vagal output by painful, frightening, or nauseating experiences. This occurs more commonly in older children and adolescents who tend to have lower blood pressure and lower resting heart rates. As the blood pressure decreases before vasovagal syncope, there is usually a prodrome of symptoms related to cerebral hypoperfusion [19]. This includes nausea, dizziness, mild tachycardia, sweatiness, and changes in vision. Situation that reduce blood pressure may further predispose a child to fainting and include prolonged standing or standing up quickly, inadequate fluid and food intake, or abnormal fluid loss from sweating in hot weather or sfter vigorous exercise.

Cardiac syncope is usually sudden and unexpected without a prodrome. It may occur during maximum exertion which is unusual for vasovagal syncope. The most likely mechanisms are reduced cardiac output from obstruction of left ventricular outflow or from atrhythrnias. Syncope related to seizures or other neurological causes may be more sudden. Seizures are often preceded by an aura

and the loss of consciousness may be accompanied by persistent tonic–clonic movements. This may be followed by residual neurological symptoms and signs as the patient awakens.

Cardiac syncope is most likely caused by obstruction of left ventricular outflow or arrhythmias. The most common LV outflow obstruction is valvar aortic stenosis, and less likely subvalvar or supravalvar stenosis. In most cases, there is a loud systolic ejection murmur associated with the obstruction that will have been detected in early childhood. An exception is hypertrophic cardiomyopathy which may only be associated with dynamic obstruction. In this case, the murmur of obstruction is a harsh systolic ejection murmur heard at the lower left sternal border and the murmur characteristically becomes louder when the patient stands or performs the Valsalva maneuver. The diagnosis may be suggested by left ventricular hypertrophy or ST-T wave changes on an electrocardiogram and it may be confirmed by echocardiography [5].

Arrhythmias that cause syncope are either ventricular tachycardia, or rarely supraventricular tachycardia at very rapid rates [19]. In these cases, the patient may recall having palpitations or chest pain just before fainting, but there is usually no prodrome of dizziness, nausea, or vision changes. Arrhythmias and syncope may occur with coronary artery and ischemic heart disease in adults, but the coronary artery abnormality in the young that would be associated with sudden collapse is the rare case of an aberrant origin of a coronary artery from the wrong sinus of Valsalva of the aortic root. This may be associated with episodic obstruction of coronary outflow, usually during exertion that causes severe chest pain, syncope, or sudden death [6].

Congenital long QT syndrome is a familial condition due to a variety of abnormalities in membrane channels within the myocardial cell that leads to ventricular tachycardia (often Torsade de pointes ventricular tachycardia) with associated syncope or sudden death. This may be precipitated by a loud noise, a sudden frightening experience, or severe emotional stress. The ECG nearly always reveals an abnormally prolonged QT interval (QTc > 470 ms) [19, 21].

Although supraventricular tachycardia (SVT) usually presents with palpitations or chest pain, it may cause dizziness and rarely syncope. At least half of SVT cases in children are related to a reentrant mechanism involving an accessory conduction pathway between the atrium and ventricle in addition to the AV node. In many of these children, a pattern of preexcitation on the ECG during sinus rhythm is present. This pattern, in association with intermittent SVT, is known as the Wolff–Parkinson–White (WPW) syndrome. The pattern includes a short PR interval and a prolonged QRS complex with a widened initial upstroke known as the delta wave [19, 21].

Complete or advanced second degree AV block may be associated with dizziness or syncope that may be sudden. Usually, children and adolescents with heart block will have been diagnosed previously and are under the care of a pediatric cardiologist. Causes include congenital heart block without underlying disease, heart block that is related to congenital heart disease or its surgical treatment, or rarely Lyme disease or acute rheumatic fever. In most cases, the diagnosis of heart block will be suspected by an inappropriately slow heart rate or by electrocardiography. Children and teenagers with permanent heart block who faint are candidates for pacemaker therapy.

Since cardiac syncope may be due to arrhythmias that are associated with underlying abnormalities on a routine 12 lead ECG, many pediatricians will obtain an ECG on any child or adolescent who faints. Other clinicians will defer the test unless the child faints a second time.

Past medical history: Children with previously diagnosed heart disease who faint unexpectedly should be brought to the attention of the pediatric cardiologist. History of previous seizures or neurological symptoms should be determined.

Family history: History of frequent fainting, fainting precipitated by a loud sound, a sudden frightening experience or severe emotional stress, or a history of unexpected sudden death in family members raises the possibility of congenital long QT syndrome, other channelopathies, or familial hypertrophic cardiomyopathy.

History

The cause of syncope (like chest pain) can usually established by taking a detailed history [19, 22]. Since the episodes are fairly discrete and relatively rare events, the physician should assume the role of a private investigator who is recreating the scene of an accident (the syncopal event). First, information about the fainting episode from witnesses should be collected. Then, recollections of the child should be carefully recorded.

The time of the day and location of the faint should be determined. Many syncopal events related to mild hypotension occur early in the morning after awakening and before the child has had any oral intake or before lunch period in school.

A complete history of the child's activities for the preceding 24 h should be reviewed for any changes in the child's well-being and activity.

Intercurrent illness, inadequate food and fluid intake or sleep, fluid loss from sweating during exercise could all predispose to a vasovagal event.

History of the symptoms immediately before the faint is crucial. Witnessing a painful, frightening or nauseous situation shortly before the faint could stimulate vagal output. The ambient temperature of the child's surroundings, whether the child was standing or just srood up at the time of the faint, and whether the child was injured during the fall should be determined.

Dizziness, nausea, or vision changes before the faint are very common with vasovagal syncope but not with syncope of cardiac or neurologic causes. Chest pain or palpitations immediately before fainting would raise suspicion of a cardiac cause.

History ofexertion at the time of the faint is crucial. Syncope during exercise should be considered cardiac in origin, although highly competitive athletes may exercise to the point of collapse even though their cardiac function is completely normal, or above normal.

Repetitive tonic–clonic movements during the faint would increase the likelihood of a seizure, although a few clonic movements may be observed after cardiac or vasovagal syncope. After a vasovagal faint, the child usually awakens quickly without neurological symptoms. Urinary or fecal incontinence, confusion, speech deficits, or paralysis after an episode of loss of consciousness would be more consistent with a seizure.

A history of previous dizziness, near syncope, and syncope should be sought. Specifically, does the child ever feel dizzy when getting up quickly from a supine position? Is the feeling similar to how the child felt before the faint occurred? If so, this would suggest that at transient decrease in blood pressure was the mechanism of the faint rather than a neurological or cardiac cause.

Physical Examination

The examination of the fainting child should include an accurate blood pressure. Postural blood pressures may be taken in the right arm when the child is supine and again after the child has been standing for 2 min. If there is a drop of 20 mmHg, systolic with standing this suggests that the child is prone to symptomatic postural hypotension. Cardiac examination in the patient with hypertrophic cardiomyopathy with obstruction may reveal a systolic ejection murmur at the left sternal border that radiates toward the upper right sternal border and to the carotids. The murmur may increase when the child stands up or performs the Valsalva maneuver. Neurological examination may be abnormal in children who lost consciousness because of a seizure.

Additional Tests

Any child who has neurological symptoms before or after the faint should probably be discussed with a neurologist. An electroencephalogram (EEG), computerized tomography (CT) scan, or magnetic resonance imaging of the head may be necessary to rule out neurological pathology. Any child who faints during exercise has palpitations or chest pain before the faint, any cardiac abnormality on examination, or a positive family history of fainting or unexpected sudden death should definitely have an ECG and probably be

referred to a pediatric cardiologist for echocardiography and other testing [19].

Anemia may predispose to fainting and should be considered in all children who faint or complain of dizziness.

In brief, most children and teenagers who faint have no preceding cardiac symptoms but have a prodrome of dizziness and nausea. There may be antecedent precipitating factors for a vasovagal event. Recovery is rapid, with no neurological sequelae. Past medical and family history is negative and cardiac and neurological examinations are normal. An ECG is within normal limits. These patients most likely have vasovagal syncope and require reassurance with no further testing.

How to Counsel Patient and Families About Syncope

In general, most children with one vasovagal event do not require any further evaluation or testing and most parents will be comfortable with this approach. Consider obtaining an ECG, especially if the child has fainted two or more times. If the ECG is abnormal, referral to a pediatric cardiologist is indicated. Even with a normal ECG, children who faint repeatedly will likely require further attention of the primary physician and may need consultation with a pediatric neurologist or cardiologist.

An approach to the child and family with vasovagal syncope is described in Table 3.1.

Acknowledge that fainting is common in older children, that it is benign in the vast majority, and that the patient is in the "fortunate majority" who faint because of a mild drop in blood pressure rather than a serious problem of the heart or nervous system. As a rule, this implies that the child has a relatively low blood pressure, which is probably a good sign for the child's long-term health. Although medical treatments are available to raise blood pressure and reduce the likelihood of fainting, they have side effects and should not be considered as initial therapy. Usually, treatment involves careful attention to prevention of predisposing causes of subsequent vasovagal fainting. The child should first be aware of the feelings that precede a faint. If these symptoms recur, the child should immediately sit down or recline since the faint will usually occur when the child is standing and will often be prevented by reclining prolonged standing or abrupt standing up, prolonged periods without cating. Prolonged periods without eating, drinking, or sleeping should be avoided. Excessive salt restriction should be avoided and good hydration should be encouraged before, during, and after exercise when muscle vasodilation and dehydration from sweating can lead to hypotension. If the patient continues to faint repeatedly, or if the history, family history, examination, or ECG suggest possible cardiac disease then consultation with a pediatric cardiologist or neurologist should be considered [19–22].

Case Study 3

A 13-year-old girl is seen for evaluation of syncope and near syncope. Two weeks ago she awakened at 6 a.m. and immediately went into the bathroom to take a shower. Toward the end of the shower she recalls feeling dizzy and nauseated. Her vision became darker. She recalls trying to get out of the shower and recalls nothing after this. Her mother heard a loud noise in the bathroom and found her daughter on the floor with a large bump on the side of her head. Her daughter began to awaken and was carried by her father to her bed where she rested for a few minutes. She was taken to the emergency department where the large head bruise was noted. The remainder of her examination was normal. An ECG and a CT scan of the head were normal.

Questions

1. The cause of her symptoms is most likely which of the following?
   (a) Arrhythmia
   (b) Cardiomyopathy
   (c) Seizure
   (d) Brain neoplasm
   (e) Vasovagal/postural hypotension

2. What would the next step be in the patient's management?
   (a) Refer to a pediatric cardiologist
   (b) Refer to a pediatric neurologist
   (c) Obtain an outpatient EEG
   (d) Obtain an echocardiogram
   (e) Reassure, instruct about preventive measures, and observe

Answers

1. (e): This is a classic history for a vasovagal faint. She had just awakened early in the morning when the heart rate and blood pressure are relatively low. She had not eaten or drunk anything before taking the shower. The warmth of the shower likely caused further vasodilatation and she was also in a standing position. In addition, the faint was preceded by symptoms of dizziness, nausea, and altered vision. These are all consistent historical features of a vasovagal faint. The event was not sudden and was not preceded by chest pain or palpitations, unlike syncope resulting from cardiac causes. There were no tonic–clonic movements noted by her parents and she had no residual symptoms after the faint, unlike a seizure. Finally, the head CT scan was normal, thus excluding most neurological causes.

2. (e): It would be reasonable to reassure, instruct, and observe this patient and her family. The history of the faint and subsequent dizziness is very consistent with a vasovagal faint. In this case, the ECG already excluded long QT syndrome, preexcitation, or severe ventricular hypertrophy. It would be reasonable to perform an ECG if this had not already been done, especially if there were any family history of arrhythmias, fainting, or sudden death.

Additional Question

3. She was discharged home without additional testing and with reassurance and education about preventive measures. Since then, she had one episode of severe dizziness when she got up quickly from the sofa in order to get a snack in the kitchen. Her vision also became dark, but she sat down again and the symptoms resolved. Her mother called to ask whether a medication could be prescribed for her daughter. What would your next step be?
   (a) Refer to a pediatric cardiologist
   (b) Refer to a pediatric neurologist
   (c) Obtain an outpatient EEG
   (d) Obtain an echocardiogram
   (e) Reassure, instruct about preventive measures, and observe

Answer

3. (e): Reinstruct the family that dizziness will occur with abrupt standing or prolonged standing and acknowledge that the patient appropriately responded to the symptom by sitting down and waiting for the dizziness to subside.

Palpitations

The sensation of the heart beating irregularly, too fast, too slow, or too hard is a common complaint in older children and adolescents [21]. It is an increasingly frequent reason for referral to the pediatric cardiologist. Often children will complain of the heart beating hard when they are actually complaining of chest pain in which case they are usually evaluated primarily for chest pain.

The most common cause of palpitations in an otherwise healthy child who complains that the heart is beating too fast is either normal sinus rhythm that the child simply senses as being too fast or sinus tachycardia that is benign. The rate of sinus tachycardia that is recorded on event recordings in symptomatic children is usually less than 160 beats per minute (bpm). The causes of sinus tachycardia (except for congestive heart failure) are usually noncardiac and include fever, pain, anxiety, infection, anemia, hypoxemia, hyperthyroidism, side effects from medications, sensitivity to caffeine or other over the counter stimulants, or rarely a catecholamine-secreting tumor (pheochromocytoma) [21].

The cardiac causes of palpitations in a child who complains of an irregular heart beat are either premature atrial or ventricular contractions that are usually benign, or rarely significant tachy- or bradyarrhythmias.

Premature atrial or ventricular contractions are benign in children with an otherwise normal cardiac history and examination. The most common significant arrhythmias causing palpitations in the young are supraventricular tachycardia, atrial flutter, and, less frequently, ventricular tachycardia. As a rule, significant arrhythmias often persist for minutes or hours and may require medical treatment to be converted to sinus rhythm. This often provides sufficient time for the child to be taken to a clinic or ED where an ECG can be performed and will establish the exact diagnosis. These arrhythmias can be episodic and rather short lived, however. In those cases, Holter or event recordings will help to confirm the patient's cardiac rhythm at the exact time symptoms occur.

The most common causes of supraventricular tachycardia (SVT) or ventricular tachycardia (VT) are the same as those which cause arrhythmias and syncope. SVT may be associated with preexcitation (e.g., WPW syndrome). VT may be associated with congenital long QT syndrome, Brugada syndrome, or hypertrophic cardiomyopathy. For this reason, a 12-lead ECG is a reasonable screening test to perform in children who complain of palpitations [21].

History

Like chest pain and syncope, palpitations should be evaluated first by taking a thorough history and performing a complete physical examination. After this, the cardiac rhythm should be defined when the child is having palpitation symptoms.

The history should include all details about the child's general health, activity, and other possible cardiac symptoms at the time of the palpitations. Any child with known cardiac disease who presents with palpitations should be referred to the child's pediatric cardiologist for further evaluation.

Inquire whether the child, child's parents or other family members, school nurse, or other health professionals have attempted to take the child's pulse when the palpitations have occurred and, if so, whether the heart rate could be determined.

History of chills with the palpitations would suggest sinus tachycardia secondary to fever or infection. History of recent blood loss or heavy periods in female adolescents would suggest anemia. Heat intolerance, unexplained weight loss, or increased emotionality are symptoms of hyperthyroidism.

A history of all medications, including over the counter medications, and intake of all energy drinks and caffeinated beverages should be obtained.

Palpitations with severe headaches, abdominal pains, pallor, or dizziness are symptoms associated with pheochromocytoma. This diagnosis will often be associated with hypertension on examination. Combinations of these symptoms could also be associated with an anxiety disorder. Other symptoms associated with anxiety disorder include restlessness, abdominal pains, flushing, muscle tension, sweating, and trembling [23, 24].

The family history should include questions about other family members with arrhythmias, WPW syndrome, or other rhythm heart rhythm problems that require medication or catheter-directed treatment. A family history of unexpected sudden death (long QT syndrome or hypertrophic cardiomyopathy) should be obtained.

Physical Examination

The physical examination of a child with palpitations who has no previous history of heart disease is usually normal. Fever, pallor, signs of dehydration or infection, hypoxemia, thyromegaly, and hypertension should be excluded since these symptoms and signs would suggest treatable, noncardiac causes of sinus tachycardia.

If an arrhythmia is detected during the examination then an ECG should be performed at that time, if possible. If the cardiac examination or ECG is abnormal, then the child should be referred to a pediatric cardiologist. If the examination and ECG are normal and there are no other predisposing causes for sinus tachycardia, then observation without intervention is appropriate.

If palpitations persist or if they are causing undue anxiety for the patient or family, then the physician should consider ordering an event

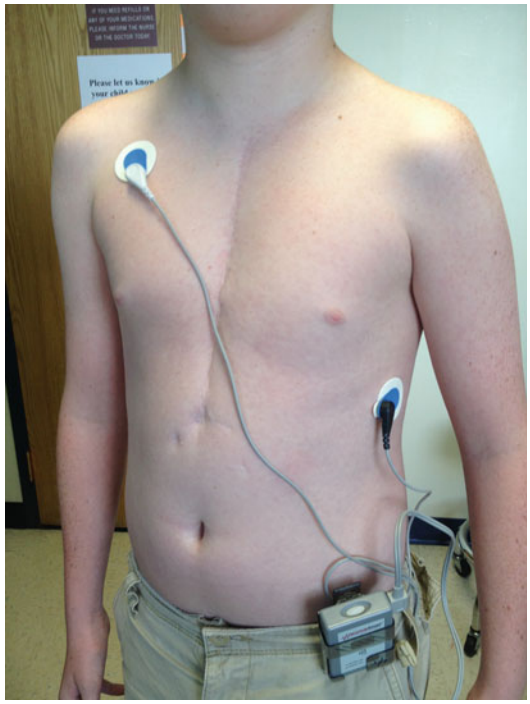

Fig. 3.1 Young adolescent post open heart operation wearing event recorder

recorder for the child (Fig. 3.1) [25]. This is a small device that is worn by the child for up to 30 days. It constantly retains the most recent 30 s of the child's heart rhythm in its memory. When the child has palpitations, either the child or parent presses a button on the device that immediately stores the preceding 30 s of cardiac rhythm until it is downloaded. This can then be transmitted via telephone to a device which produces an ECG rhythm strip from the digitally stored cardiac rhythm. This provides a recording of the cardiac rhythm at the exact time of the child's palpitations.

When palpitations become troublesome, the event recordings of cardiac rhythm during symptoms can provide the best reassurance that the palpitations are not due to a significant cardiac arrhythmia. If the recordings do reveal a significant arrhythmia, then the child should be referred to a pediatric cardiologist. If the recordings reveal premature atrial or ventricular contractions alone, some physicians might simply reassure the patient and others would refer the child to a pediatric cardiologist simply to exclude the presence of any underlying heart disease before providing reassurance that the premature beats are completely benign. If the recordings reveal normal sinus rhythm or sinus tachycardia, then causes of sinus tachycardia should be considered. If the evaluation is otherwise normal, then reassurance and reduction of predisposing factors (i.e., caffeine intake) are all that is necessary.

How to counsel the parents and patient with palpitations that the palpitations are benign or "functional."

The family of a child with palpitations that are associated with normal sinus rhythm or mild sinus tachycardia should be reassured that palpitations are a relatively common complaint in children and adolescents and that they are usually benign. The child is in the "fortunate majority" of children with a benign explanation for the palpitations. As with discussions of chest pain and syncope, Table 3.1 summarizes an approach to counseling the child and family in this circumstance. If the palpitations are very infrequent, then reassurance and perhaps a follow-up visit in 6–12 months may be considered. Options to reevaluate the child if symptoms become more bothersome should be offered. If an event recording has not been done, explain that this may be helpful in the future to evaluate the palpitations more accurately if they become more frequent or are of longer duration. If an event recording has been performed and demonstrates no arrhythmia during the child's symptoms, then parents and child can be reassured that the palpitations are not related to a significant heart condition.

The usual cause of palpitations is mild sinus tachycardia, a mild, natural increase in heart rate that occurs when a person is exercising or excited. Sometimes, the heart rate will increase in growing children for no apparent reason. In these cases, we attribute this to hormonal effects or simply to wider ranges of normal heart rates that younger children have.

Mild sinus tachycardia is a known side effect of medications used to treat attention deficit hyperactivity disorder (ADHD) [26, 27]. In the case of a child with palpitations who is also taking a medication to treat ADHD, an ECG is advisable. If the palpitations are frequent or dis-

ruptive, an event recorder will document the cardiac rhythm at the time of the child's symptoms. If sinus tachycardia is documented, a discussion with the family and perhaps the child's school may be necessary. The parents should be reassured that mild sinus tachycardia is not harmful to the heart. If the child's ADHD is well treated with the medication, then the family and child may accept occasional palpitations resulting from the medication in order to maintain the medication's positive behavioral effects. Adjustment of the dosage of the medication, or change to a different medication may also lead to relief of symptoms.

Case Study 4

A 10-year-old boy is seen in your office because of complaints of palpitations and chest discomfort. Over the last month or so, he has complained to his mother, "Mommy, feel my heart! It's beating fast!" His mother will put her hand on his chest and the heart appears to be beating faster than normal. He says that his heart "hurts" when this happens, but cannot be more specific about this. The episodes happen both at rest or when he is playing, but usually when he is resting. His mother has told him to sit down, rest, and try to relax and this seems to help him. The palpitations last no more than 10 min. Of note, he was diagnosed with ADHD and was placed on a medication for this about 2 months ago. Since then, his school performance has improved significantly.

Question

1. What further information could be most helpful in your management of this child?
   (a) Further detail about academic performance
   (b) Which ADHD medication he has been prescribed and its dosage
   (c) His ability to exercise
   (d) Presence of a cardiac murmur
   (e) Further detail from mother regarding his episodes

Answer

1. (e): This is tricky question. Many parents are capable of taking a radial or carotid pulse. If possible, the parents should be enlisted to do this, if they have not already attempted to do so. Most tachyarrhythmias are associated with heart rates greater than 160 bpm in children. If the parent appeared reliable and reported that the child's heart rate was 120 bpm, then this is likely to be sinus tachycardia and benign.

Additional Question

2. The child's mother had taken his pulse and it was 120–140 bpm. He was seen by the school nurse because of palpitations, and she detected a pulse of 134 bpm with a normal blood pressure. Initial evaluation was normal and reassurance was provided. He had repeated episodes, however, occurring 2–3 times per day for several weeks and his mother brought him back to you for reevaluation. His medication dose is unchanged, and he is not taking any caffeine-containing beverages or other over the counter medications. Physical examination is again normal. An ECG is normal. What would you do next?
   (a) Reassure
   (b) Perform thyroid function tests
   (c) Check urine catecholamines
   (d) Obtain an echocardiogram
   (e) Review management of ADHD

Answer

2. (e): The child and mother have returned to your office and reassurance may be difficult. Hyperthyroidism and pheochromocytoma are causes of palpitations in children, but they are very rare and the child's palpitations began not long after the ADHD medication was started. The child's dose of medication should be reviewed. Perhaps a small decrease in dosage may preserve its beneficial effects but reduce his palpitations.

Additional Question

3. The child continues to have palpitations in spite of a reduction of his stimulant dosage. One episode lasted 30 min and his mother had difficulty counting his pulse at that time. His examination including blood pressure is normal. You ordered an event recording during which an ECG was captured during six different episodes of palpitations. All revealed mild sinus tachycardia, rate 120–130 bpm. The child is having difficulty in school again and is complaining of intermittent headaches and abdominal pain. What would be your next step?
   (a) Reassure
   (b) Consider anxiety disorder
   (c) Check urine catecholamines
   (d) Obtain an echocardiogram
   (e) Begin beta blocker

Answer

3. (b): Answer c would be acceptable, although pheochromocytoma is relatively rare, and his blood pressure has been normal. His persistent symptoms will unlikely improve with simple reassurance. An echocardiogram is unlikely to uncover a cardiac cause in the absence of abnormalities on examination or previous ECG. Beta blocker would only be considered in extreme cases. This child is having several symptoms of an anxiety disorder. Further psychological evaluation and treatment should be pursued.

Conclusions

Chest pain, syncope, and palpitations in children usually represent benign conditions, do not require aggressive treatment, and may be considered to some degree "functional." Careful evaluation of these symptoms including obtaining of a history in a nonbiasing fashion, and appropriate reassurance of the patients and their families can help avoid unnecessary testing and referrals. Some of the affected children may benefit from referral for mental health therapy.

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Functional Symptoms in Pulmonology: Taking Your Breath Away

4

Ran D. Anbar

Abstract

Children with respiratory symptoms often are affected by a functional contribution to their presentation. In order to provide appropriate comprehensive care, clinicians need to identify and address these issues in such children. Some respiratory diagnoses are recognized generally to be of functional origin including functional dyspnea, habit cough, and vocal cord dysfunction. The criteria for establishing these diagnoses are discussed in the first section of this chapter. However, commonly unrecognized are the children with diagnosed physiologic disease such as asthma, bronchopulmonary dysplasia, or cystic fibrosis who develop additional or worsening symptoms due to emotional or psychosocial overlay. Whether triggered by physiological or psychological factors, symptoms may be identical, and thus clinicians may even be unaware of the psychological impact on a patient. The second section of this chapter will address how clinicians might recognize and offer optimal treatment for such patients.

Keywords

Anxiety • Asthma • Dyspnea • Habit cough • Vocal cord dysfunction

Children who present with respiratory symptoms often are affected by a functional contribution to their presentation. In order to provide appropriate comprehensive care, clinicians need to identify and address these issues when they exist [1].

Some clinicians will readily recognize the functional origin of certain respiratory diagnoses including functional dyspnea, habit cough, and vocal cord dysfunction (VCD). The criteria for establishing these diagnoses are discussed in the first section of this chapter.

However, less commonly recognized are children with diagnosed physiologic disease such as asthma, bronchopulmonary dysplasia, or cystic fibrosis (CF) who concurrently develop symptoms arising as a result of emotional or psychological issues. Sometimes, these symptoms are identical irrespective of their origin and clinicians may

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even be unaware of the psychological impact the physical illness and its consequences has on a patient. The second section of this chapter addresses how clinicians might recognize and offer optimal treatment for such patients.

Commonly Recognized Functional Respiratory Disorders

Functional Dyspnea

Clinical Presentation

Dyspnea can be defined as difficult or labored breathing or shortness of breath. It can occur in the course of normal exercise, in the context of cardiopulmonary disease, and/or because of a psychological reaction. Functional dyspnea can present with rapid or noisy breathing, or may solely be evident to the patient [2]. Clinical examples of functional dyspnea include hyperventilation, sighing dyspnea, and VCD (discussed below) [3].

Patients with functional dyspnea frequently report that they feel as though they cannot inhale fully or catch their breath. When exercise is a trigger of functional dyspnea, patients report development of symptoms much earlier during exertion than would be expected based on their level of conditioning [2]. Dyspnea resulting from hyperventilation often is associated with symptoms of hypocarbia including dizziness, chest wall pain from muscle spasm, and tingling or numbness of their extremities (paresthesia). Patients with panic disorder frequently report an intense fear of dying when they first develop associated dyspneic episodes [2].

In the absence of a concurrent medical illness, the physical examination of most patients with functional dyspnea is normal, with the exception of rapid and deep breathing during an acute hyperventilation episode. Spirometric testing at rest also is normal. Exercise pulmonary function testing can help establish whether the dyspnea is related to an underlying physiologic abnormality or normal physiologic limitation [4]. Hyperventilation provocation tests with concurrent measurements of end-tidal carbon dioxide have been utilized to diagnose patients with hyperventilation. In comparison to unaffected individuals, patients who hyperventilate demonstrate lower levels of carbon dioxide at rest, and a slower recovery of their carbon dioxide level following voluntary overbreathing [5].

Natural History

Functional dyspnea is well known to be related to psychological issues. Patients with hyperventilation frequently have been diagnosed with anxiety, panic, conversion disorder, and phobic symptoms [6, 7]. Further, the majority of patients with generalized anxiety and panic disorders develop hyperventilation [8].

Differential Diagnosis

As dyspnea often is the result of cardiopulmonary disease, clinicians must be cautious in making a diagnosis of functional dyspnea and consider a large differential diagnosis [2]. For example, dyspnea can occur as a result of an upper airway obstruction such as from a foreign body, angioedema, or an infectious process. Common lower airway disease that causes dyspnea includes asthma, laryngotracheobronchitis (croup), and pneumonia. Cardiac causes include arrhythmias and congestive heart failure. Other diagnoses to consider include anemia, metabolic alkalosis, and toxigenic causes such as carbon monoxide poisoning, or salicylate overdose [2]. An appropriate workup for dyspnea that is negative can serve as the basis for reassurance as therapy.

Functional dyspnea typically resolves within minutes without specific therapy, in contrast to dyspnea that is the result of cardiopulmonary or metabolic disease [2].

Treatment

Functional dyspnea can improve with reassurance [7], acupuncture ([9], Chap. 23), biofeedback ([10], Chap. 20), breathing techniques [11, 12], cognitive behavioral therapy (Chap. 19), and hypnosis ([13], Chap. 21).

Habit Cough

Clinical Presentation

Children with habit cough typically present with a loud, disruptive cough that has been described

as "honking," "brassy," or "barking." The cough often increases when patients pay attention to it, usually improves when children are distracted (e.g., while playing a videogame), and ceases when they are asleep. Notably, during the process of falling asleep habit cough sometimes intensifies [14–16]. (Video 4.1 shows a patient with habit cough, who is receiving treatment with self-hypnosis.)

Atypical forms of habit cough include children in whom the cough is not disruptive and who manifest some coughing while they are asleep [14].

The physical examination of children with habit cough usually reveals no abnormalities [16]. Pulmonary function testing of children with habit cough is normal [14, 16], although if the cough has been triggered in association with another pulmonary disease such as asthma, the testing may reflect abnormalities associated with the other disease. Other diagnostic testing often is unnecessary in a child with classic habit cough, when the diagnosis can be made based on the characteristic clinical presentation. Chest radiography studies are normal [16]. Bronchoscopy usually is normal, although sometimes patients develop tracheal petechiae as a result of their intense coughing, which should not be confused as a primary trigger of the cough [1]. In other patients, localized tracheomalacia has been identified occasionally, which was thought to have been preexisting in patients who later developed a habit-like cough [17].

Natural History

Habit cough often is triggered by upper respiratory infections, but persists once the infection resolves. It also has been reported to be triggered by asthma [14].

In one case series, half of children with habit cough missed at least a week of school as a result of the disruptive nature of their symptom [14].

Without therapy, habit cough has been reported sometimes to persist for years [18].

Differential Diagnosis

The differential diagnosis for habit cough includes asthma, protracted bacterial bronchitis, pertussis (whooping cough), and tracheomalacia [16]. While habit cough tends to cease when patients are asleep, cough attributable to asthma, bronchitis, and pertussis typically persists during sleep. Tracheomalacia can cause a loud, disruptive cough, which can mimic the type of cough heard in patients with habit cough. However, the cough associated with tracheomalacia often occurs while patients are asleep and does not typically change in frequency when patients are distracted as does habit cough [16].

Treatment

Effective therapy for habit cough includes psychotherapy (see Chap. 19), hypnosis ([14], Chap. 21), and suggestion therapy [15, 16]. During the latter therapy, over a few minute period of time, it is suggested repeatedly to patients that they can control their urge to cough, while they are sipping small amounts of room temperature water or breathing nebulized lidocaine (0.5 ml of 1 % lidocaine diluted to 3 to 5 ml with normal saline).

Vocal Cord Dysfunction

Clinical Presentation

Patients with VCD typically present with complaints of difficulty with inhalation and occasional associated stridor. Frequently, they report a feeling of blockage of their airway at the level of the neck or upper chest [19]. Some patients also develop symptoms suggestive of associated hyperventilation including dizziness, paresthesia, and tremors [2].

VCD frequently is triggered by athletic activity in teenagers and resolves within several minutes of rest. It is less common for VCD to occur during rest [19]. On rare occasions VCD has been reported to persist during sleep [20].

VCD usually occurs as the result of adduction of the vocal cords during inhalation, although on infrequent occasions the adduction occurs during exhalation as well or only during exhalation [19, 21]. Direct visualization of the vocal cords during a VCD episode is diagnostic of this condition. Dynamic 320-slice computerized tomography (CT) also may be helpful in the diagnosis of VCD [22]. Indirect evidence consistent with VCD

includes presence of inspiratory stridor during the physical examination and flattening of the inspiratory flow loops during spirometry [4, 19].

Natural History

VCD often occurs in association with stress. The common type of VCD that is induced by exercise (EIVCD) often occurs in high achieving teenagers (more likely to be female) [23], who are stressing themselves greatly in order to excel. Thus, EIVCD is more likely to occur during athletic competitions rather than practice. VCD has been noted to sometimes occur in association with psychosocial stressors such as parental divorce, academic difficulties, or mental, physical, or sexual abuse of the patient [20, 24].

Differential Diagnosis

Patients with VCD often are first diagnosed as having asthma because of their complaints of shortness of breath. The diagnosis is further complicated by the fact that VCD has been reported to occur concurrently in many patients with asthma (6 % in one series) and exercise induced bronchospasm (31 %) [23]. VCD can be differentiated from bronchospasm given its associated stridor and upper airway obstruction as opposed to the wheezing and lower airway obstruction evident in bronchospasm. Other diagnoses in the differential include exercise-induced laryngomalacia, vocal cord paresis or paralysis, or subglottic stenosis [19]. All of these diagnoses can be excluded based on laryngoscopic evaluation of the upper airway. VCD has been reported to occur in some patients with Chiari I malformation, and this possibility should be considered in very young children with VCD [25], or if patients' symptoms do not resolve with therapy that typically helps resolve VCD (see below).

Treatment

Biofeedback (Chap. 20), breathing techniques [12, 26], hypnosis (Chap. 21), and speech therapy [27] have been utilized successfully in the treatment of VCD. Patients with EIVCD frequently respond to treatment with an inhaled anticholinergic medication that is administered prior to exercise [19, 28] Chap. 26.

When Respiratory Disorders Are Complicated by Functional Symptoms

Clinicians should suspect that a functional component may be affecting the presentation of children with diagnosed respiratory disorders when their symptoms do not improve as expected with standard medical therapy, or if their symptoms cause more distress than typically observed. Further, emotional reactions related to the respiratory symptoms increase the probability of a significant functional component. Table 4.1 lists symptoms that frequently occur in patients with respiratory symptoms that arise as a result of functional issues. It should be noted that emotional issues such as anxiety may be the result of a primary disorder or have arisen in reaction to the medical condition [1].

The medical community often fails to consider functional symptoms as a major cause of respiratory symptoms that fail to respond to standard therapy. Reasons for this include the physician's lack of expertise with recognizing functional complaints and their treatment [28]. Nonetheless, such complaints are very common. For example, in clinical practice at a tertiary care Pediatric Pulmonary Center of 301 consecutive children who were referred for assessment of their respiratory symptoms, 20 % of 6–11-year-olds, and 31 % of 12–18-year-olds were diagnosed as having a functional component that was a primary cause of their presentation [29].

When a functional contribution is suspected, clinicians should reconsider concurrent assessment and management of organic, psychological, and psychosocial issues. Otherwise, patients may end up undergoing unnecessary diagnostic tests and receiving ineffective medications, which can cause further complications. Further, extensive medical testing in response to unresolved symptoms may convince patients that they have a medical condition that the clinician has failed to diagnose. Such a conviction may make patients more resistant to the idea that some of their symptoms are functional in nature [1].

Table 4.1 Symptoms suggestive of functional respiratory disorders

Respiratory symptoms
Chest Pain in the absence of cardiac or gastrointestinal disease
Difficulty with inspiration[a]
Disruptive cough[b]
Dyspnea despite normal lung function[c]
Hyperventilation (which patients may term breathing too fast)
Inspiratory noise (e.g., stridor, gasping, rasping, or squeak)[a]
Localization of breathing problem to the neck or upper chest[a, c]
Sighing[c]
Other symptoms
Anxious appearance
Dizziness[c]
Feeling something is stuck in the throat Lifted shoulders
Palpitations
Paresthesias[c]
Shakiness[c]
Tics[b]
Weakness
Symptom characteristics
Absence during sleep or when patient is distracted
Associated with a particular location or activity
Emotional response to symptoms
Emotional trigger of symptoms
Exposure to traumatic life event
Incomplete response to medications

Adapted from Anbar RD, Geisler SC. Identification of children who may benefit from self-hypnosis at a pediatric pulmonary center. BMC Pediatrics 2005; 5: 6. With permission from BioMed Central Ltd

[a]Suggestive of vocal cord dysfunction

[b]Suggestive of habit cough

[c]Suggestive of functional dyspnea/hyperventilation

There is an extensive literature reporting the association of childhood respiratory disease with psychological issues that can lead to the development of functional symptoms. Additionally, behavioral problems can affect adherence to the prescribed therapeutic regimen for an organic respiratory disorder, and thus be associated with worsening respiratory symptoms [30].

For example, asthma exacerbations are known to occur as a result of stressful life events [31, 32]. Pediatric patients with asthma frequently are found to have clinical anxiety [33, 34]. In turn, anxiety and depression have been shown to be associated with increased asthma symptoms [35]. Patients with CF are at increased risk of developing adjustment problems and internalizing symptoms of depression and anxiety. Thus, children with CF may report physical symptoms associated with psychological difficulties such as abdominal discomfort, muscle tension, shortness of breath, and/or tremors (e.g., as a result of anxiety) [30]. Patients with CF who have developed concurrent attention deficit or oppositional defiant disorders have been reported to be less adherent to their therapies, which has been associated with a poorer prognosis [36]. Patients who were born prematurely and developed bronchopulmonary dysplasia also have been reported to be at increased risk of developing anxiety and attention disorders in later childhood [37, 38], which may affect the nature of their respiratory symptoms.

Treatment

When it is suspected that a respiratory symptom is complicated by a functional problem, patients can be offered an appropriate intervention based upon their developmental age, which is directed at either improving the underlying stressor or teaching the patient how to control their symptoms [1].

For example, if a patient with asthma develops shortness of breath while taking tests at school, the first step is to help the patient understand that his respiratory distress is related to anxiety rather than an asthma exacerbation. To help relieve the anxiety, the patient and his family might be encouraged to discuss the problem with school officials and the patient might be given appropriate coaching and accommodations so that test-taking causes much less stress. Alternatively, the patient might be taught self-regulation strategies to calm himself while taking examinations. Either strategy could help resolve the episodes of shortness of breath.

Interventions that can help improve the underlying stressor include cognitive behavioral therapy (Chap. 19), play therapy (Chap. 28), and reassurance.

Interventions that can help the patient calm himself include meditation (Chap. 24), self-hypnosis (Chap. 21), and yoga (Chap. 25).

Interventions that can help calm the patient but are dependent for the long term on equipment or a healthcare provider include acupuncture (Chap. 23), biofeedback (Chap. 20), and guided imagery (Chap. 22).

There is no evidence in the literature comparing the efficacy of the various aforementioned interventions for functional respiratory disorders. Interventions for individual patients may be chosen based on their availability (e.g., at the office of the primary care provider vs. requiring referral) and patient preference.

For patients whose psychological stress has been caused solely by their reaction to an organic disease, treatment of the underlying pulmonary condition can lead to resolution of the associated functional symptoms [1]. For example, a patient with asthma may become increasingly anxious with associated hyperventilation whenever his asthma is under poor control. When the patient's asthma is brought under control, his anxiety and associated functional symptoms resolve.

Caveats

In some cases, functional respiratory symptoms can cause physical pathology. For example, severe habit cough has been reported to cause airway petechiae or rib fractures, severe VCD has led to loss of consciousness, and hyperventilation has triggered bronchospasm. It is important that clinicians not confuse secondary organic changes in such situations as the primary causes of the patient's presentation [1].

Another common mistake made by clinicians is to assume that abnormalities identified during medical investigations necessarily are the cause of patients' symptoms. For example, a patient with inspiratory stridor as a result of VCD may be found to have an abnormal methacholine challenge test that is diagnostic of asthma. A clinician may thus treat this patient for asthma, even though the abnormal test in actuality was an incidental unrelated finding. When the patient fails to respond to this therapy, the patient erroneously may be characterized as having "difficult" asthma [39].

Some patients develop persistent functional symptoms as a result of a significant psychological disturbance [24]. For example, a patient may develop habit cough in association with depression related to her parents' divorce, or VCD as a result of a conversion disorder related to sexual abuse. Such patients will not respond easily to self-regulation strategies as described above and should be referred for evaluation by a mental health professional (Chap. 18). In rare occasions, patients may benefit from pharmacotherapy for their psychiatric condition such as the use of antidepressants (Chap. 26) [28].

Finally, misdiagnosis of an organic disease as being caused by a functional respiratory symptom can delay appropriate medical treatment. Thus, clinicians should reevaluate their diagnosis of a functional disorder if patients' symptoms fail to improve with therapy that typically helps resolve such conditions [1].

Case Studies

Case 1: Asthma [39]

A 9-year-old boy presented with a long history of recurrent cough and wheezing in association with upper respiratory infections, exercise, change in weather, and exposure to allergens. In later childhood, the child often awoke at night due to cough, even when he was otherwise well. Sometimes, he developed cough and difficulty breathing in association with strong emotions, such as anger or sadness, or when he laughed or cried. His symptoms improved with use of bronchodilator therapy, but use of daily inhaled fluticasone, montelukast, and weekly allergy immunotherapy did not resolve episodic recurrence of his symptoms. As a result, the patient required multiple courses of oral steroids and several hospitalizations for treatment of severe and refractory respiratory exacerbations. He had a history of occasional regurgitation of food, and posttussive emesis. He also suffered from recurrent otitis media and sinusitis until he underwent placement of bilateral tympanostomy

tubes at the age of 4 years, after which he no longer developed otitis. The patient had a history of possible sexual abuse in infancy. His parents divorced when he was a year old. When clinically well, his physical examination was normal. A pulmonary function test revealed a mild obstructive pattern with a significant improvement following bronchodilator administration.

Questions

1. What is the patient's likeliest primary diagnosis?
   (a) Asthma.
   (b) Gastroesophageal reflux.
   (c) Habit cough.
   (d) Immunodeficiency.
   (e) Malingering.
2. All of the following may explain why the patient continued to develop wheezing episodes despite treatment with chronic inhaled corticosteroid or montelukast therapy EXCEPT:
   (a) Lack of adherence to his prescribed therapy.
   (b) His asthma management is complicated by gastroesophageal reflux.
   (c) His asthma management is complicated by emotional triggers.
   (d) Steroid resistance.
   (e) His asthma was severe
3. Given the child's lack of asthma control despite preventive therapy, what is the best next step in this patient's management?
   (a) Review with the patient and his family about the importance of adherence to prescribed therapy.
   (b) Begin therapy with antireflux medications.
   (c) Teach the patient better ways of dealing with his emotions.
   (d) Treat the patient with mometasone rather than fluticasone.
   (e) Start the patient on chronic every-other-day oral methyl prednisolone.

Answers

1. (a): The patient's cough, wheeze, and difficulty breathing in association with upper respiratory infections, exercise, and emotions are characteristic of asthma. That diagnosis is supported by the history of responsiveness to bronchodilator therapy and characteristic pulmonary function test results. While gastroesophageal reflux may complicate the management of asthma [40], it cannot be considered the primary diagnosis given that the triggers of this patient's respiratory symptoms do not typically aggravate gastroesophageal reflux. Patients with habit cough classically do not demonstrate abnormal pulmonary function [16]. As the patient's otitis media episodes resolved once he underwent tympanostomy tube placement and because he has had no other major bacterial infections, it is unlikely he has an immunodeficiency. Malingering is a rare diagnosis and typically is not triggered by infections or exercise [1].
2. (d): As the patient responded to systemic steroid therapy, it is unlikely that he had steroid resistance. Further, it is unusual for a patient with severe and steroid-resistant asthma to have a mild obstructive pattern on pulmonary function testing.
3. (c): This patient had received a number of preventive asthma medications without improvement. Thus, changing the inhaled steroid was unlikely to help. If not previously and repeatedly discussed, review of the importance of adherence could be helpful. Given the severe nature of this patient's asthma, it is likely that the symptoms compatible with gastroesophageal reflux were the result rather than the cause of his uncontrolled asthma. Chronic systemic steroid therapy comes with notable toxicity and should be considered only as a last step when all else fails. When this patient was taught self-hypnosis in order to promote self regulation, calm himself and modulate his emotional responses, his asthma symptoms resolved within a day, thus demonstrating that for him, there was a strong emotional component to his presentation significantly affecting the underlying asthma.

Case 2: Cough [41]

A 12-year-old boy developed an upper respiratory infection with an associated loud, honking cough that persisted during the days for several

weeks, but resolved when he was asleep. He had a history of coughing for up to 2 weeks following upper respiratory infections, which typically he would contract three times a year. He had no history of wheezing or shortness of breath. The boy reported an associated burning in his throat, but no stomachaches, heartburn, regurgitation, or emesis. He has not suffered from recurrent otitis media, sinusitis, bronchitis, or pneumonia. His physical exam and pulmonary function testing results were normal. Use of prednisone, hydrocodone, metoclopramide, omeprazole, and over-the-counter cough suppressants was not of benefit. Because of the disruptive nature of the cough, the patient had missed 50 days of sixth grade and received home tutoring. Nonetheless, he was able to maintain a high grade point average. He said he missed his friends and wanted to return to school.

Questions

1. What is the likeliest diagnosis for this patient's current cough?
   (a) Asthma.
   (b) Gastroesophageal reflux.
   (c) Habit cough.
   (d) Immunodeficiency.
   (e) Malingering associated with school avoidance.
2. Which of the following is an appropriate next step in the management of this patient?
   (a) Reassurance of the patient that the cough is not the result of a medical condition and will resolve on its own.
   (b) Instruction in self-hypnosis utilizing imagery to help the patient relax.
   (c) Provision of suggestion therapy wherein the patient is reassured repeatedly that his cough can be controlled.
   (d) Use of oral benzonatate (Tessalon Perles).
   (e) B or C.
3. If the cough does not resolve immediately after therapy is initiated, should the patient be told that he must return to school no matter how the disruptive the nature of the cough?
   (a) No. The stress of the on-going cough in a social setting would serve to perpetuate it.
   (b) No. It is inappropriate to subject his classmates to the disruptive cough.
   (c) Yes. The patient will feel better when he sees his friends, and this could help resolve his cough.
   (d) Yes. Missing school as a result of habit cough often represents a benefit for patients (i.e., a secondary gain) and thus elimination of the benefit helps promote its resolution.
   (e) A or B.

Answers

1. (c): The patient's cough is characteristic of habit given its loud and disruptive nature, and its absence while he has slept [14]. While his prolonged cough after upper respiratory infections can be consistent with asthma, he does not have other symptoms suggestive of asthma, and he did not improve with use of prednisone that typically resolves an asthma exacerbation. He does not report symptoms suggestive of clinically significant gastroesophageal reflux. He does not report recurrent respiratory infections that would be suggestive of the possibility of an immunodeficiency. Malingering is a rare diagnosis.
2. (e): Either hypnosis or suggestion therapy have been reported to help resolve habit cough [16]. Habit cough can persist for years without intervention, and thus reassurance alone would not be appropriate [18]. Benzonatate is not effective in the treatment of habit cough [16].
3. (d): It is often necessary to arrange appropriate accommodations with the school in order to minimize disruption of the patient's normal routine while avoiding secondary gain derived from being absent from school [1, 14]. For example, the patient should be allowed to spend time at the school nurse's or administrative office, if necessary. Also, school staff should be informed that the cough is not a result of a communicable infection.

Case 3: Dyspnea [42]

An 11-year-old boy presented with a 4-year history of developing shortness of breath while playing soccer. He explained that his dyspnea was associated with difficulty with inhalation, causing

a loud inspiratory noise, and subsequently, a feeling of tightness and burning in the chest, tachycardia, and occasional fear. He denied associated headaches, stomachaches, nausea, or paresthesia. His breathing difficulties kept him from keeping up with his friends. In association with upper respiratory infections, this patient tended to cough for "weeks." He had a history of recurrent bronchitis and pneumonia in early childhood, but not in recent years. He underwent surgery for recurrent sinusitis at 10 years in the hopes that this would reduce his frequency of headaches, which occurred 2–3 times per week. However, his headache frequency did not change following the surgery. His physical examination and pulmonary function testing were normal. His shortness of breath persisted despite trials of several medications for asthma, allergy, and gastroesophageal reflux.

Questions

1. Of the following diagnoses, what is the likeliest cause of this patient's shortness of breath?
   (a) Asthma.
   (b) Gastroesophageal reflux.
   (c) Habit cough.
   (d) Malingering.
   (e) Vocal cord dysfunction.
2. Which of the following tests is likeliest to provide definitive evidence that his patient has VCD?
   (a) Bronchoscopy.
   (b) Chest computerized tomography.
   (c) Laryngoscopy.
   (d) Methacholine challenge test.
   (e) Spirometry.
3. Which of the following is an appropriate therapy for this patient?
   (a) Hypnosis.
   (b) Inhaled ipratropium bromide prior to exercise.
   (c) Speech therapy.
   (d) A and C.
   (e) A, B, and C.

Answers

1. (e): The patient's difficulty with inhalation and loud inspiratory noise (i.e., stridor) is suggestive of an upper airway process. Therefore, asthma is unlikely. While gastroesophageal reflux may lead to upper airway irritation, typically it is not associated with the development of tachycardia or fear. A diagnosis of VCD would be consistent with this patient's upper airway symptoms. His associated anxiety symptoms could be the result of the same psychological stress or that triggered his VCD. Cough is not a prominent part of this patient's presentation, and thus habit cough is not likely. Malingering is a rare diagnosis.
2. (c): In order to diagnose VCD, symptoms must be present. While upper airway laryngoscopy in the presence of dyspnea could demonstrate vocal cord adduction consistent with VCD [19], the anesthesia or sedation required for bronchoscopy would tend to resolve vocal cord findings, and thus prevent the clinician from establishing the diagnosis. A chest CT scan would not demonstrate vocal cord adduction. A methacholine challenge would demonstrate if a patient has hyperreactive airways, a finding not diagnostic of VCD. Finally, if the patient is symptomatic, spirometry might demonstrate blunting of the upper airway flows, consistent with nonspecific upper airway obstruction, which can be attributable to a number of different diagnoses.
3. (e): All of these therapies can be effective for the treatment of exercise induced VCD. VCD that occurs without being triggered by exercise does not improve with use of ipratropium bromide.

Conclusions

Given the high prevalence of functional symptoms, clinicians should consider the possibility of such symptoms in every child who presents with respiratory complaints. When a functional contribution is suspected, clinicians should consider concurrent assessment and management of organic, psychological, and psychosocial issues. Otherwise, patients may end up undergoing unnecessary diagnostic tests and receiving ineffective medications, which can cause further complications.

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Functional Symptoms in Gastroenterology: A Punch to the Gut

5

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Abstract

Functional gastrointestinal disorders (FGIDs) are highly prevalent in the USA and worldwide among children. These conditions can be broadly divided into those found in specific age groups as well as by symptom (e.g., nausea, vomiting, diarrhea, and abdominal pain). Collectively, these conditions account for a significant percentage of general and subspecialty practitioner visits. When unrecognized and/or untreated, these conditions can lead to significant morbidity including psychological and psychosocial disability for both patients and families. Furthermore, FGIDs are increasingly being recognized among patients with organic intestinal disorders such as Inflammatory Bowel Disease (IBD) and Celiac disease and affect disease-related management and utilization of healthcare resources. The Rome Foundation has identified key diagnostic criteria for these disorders. A majority of these disorders can be diagnosed on clinical grounds based on detailed and pertinent history and physical exam. Combined assessment of symptoms, illness experience, and psychosocial factors can be utilized to develop effective individualized treatment plans.

Keywords

Functional gastrointestinal disorders • Rome III criteria • Irritable bowel syndrome • Functional dyspepsia • Functional abdominal pain • Abdominal migraine • Functional constipation • Encopresis • Functional dysphagia • Globus • Chronic idiopathic nausea • Aerophagia • Functional diarrhea • Rumination syndrome • Inflammatory bowel disease • Celiac disease

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Abbreviations

5-HT	5-Hydroxytryptophan
AP	Abdominal pain
CCK	Cholecystokinin
CD	Crohn's disease
CIN	Chronic idiopathic nausea
COMT	Catechol-O-methyltransferase
FAP	Functional abdominal pain
FD	Functional dyspepsia
FGID	Functional gastrointestinal disorder
FOBT	Fecal occult blood test
GERD	Gastroesophageal reflux disease
GPCR	G-protein-coupled receptor
HPA	Hypothalamic pituitary adrenal
IBD	Inflammatory bowel disease
IBS	Irritable bowel syndrome
IL	Interleukin
NSAID	Nonsteroidal anti-inflammatory drugs
PI	Postinfectious
PPI	Proton pump inhibitor
SES	Socioeconomic status
SIBO	Small intestinal bacterial overgrowth
TNF	Tumor necrosis factor
UC	Ulcerative colitis

Background/Literature Review

Epidemiology

Functional gastrointestinal disorders (FGIDs) affect an estimated 25 million Americans and 5–15 % of the population worldwide [1, 2]. These disorders lead to over $200 billion dollars in costs worldwide [3, 4]. Pediatric patients with FGIDs accrue high healthcare utilization costs in part as a result of their family's search for a cause of their symptoms [5, 6] mainly for fear of missing a serious condition [7]. In turn, pediatricians order multiple diagnostic tests in search of an organic diagnosis [8]. FGIDs account for 2–4 % of general pediatric office visits and over half of pediatric gastroenterology consultations [9]. FGIDs typically have onset at ages 4–6 years and early adolescence [10–12], and their rates decline with age [13–15].

Since a significant proportion of patients with FGIDs do not seek health care, it is difficult to define the true prevalence of FGIDs in the community [16, 17]. The majority of studies in FGIDs are hospital based [18–20]. The other factor involved in accurately assessing the prevalence of FGIDs is their definition [21]. The Rome Criteria for the diagnosis of FGIDs were developed through the consensus of international experts and have undergone several iterations. The most recent set, Rome III, were developed in 2006. The disorders are subdivided into categories and classified based on predominant symptom within the pediatric age group [21].

FGIDs can be broadly divided into those found in specific age groups as well as by symptom (e.g., nausea, vomiting, diarrhea, and abdominal pain). Specific disorders addressed in this chapter are discussed in detail below (in the section entitled "The Pediatric FGIDs"). The two most common FGIDs for which patients are referred to American pediatric gastroenterologists are functional constipation and Functional Abdominal Pain (FAP) [22]. Outside the USA of the pediatric abdominal pain predominant FGIDs, Irritable Bowel Syndrome (IBS) is the most common, followed by FAP and Functional Dyspepsia (FD) [13]. Gender differences in FGIDs occur around puberty [11, 23], wherein a female predominance typically develops [13, 17, 24–26] and may be due to differences in hormonal profiles [27].

FGIDs can cooccur with extraintestinal somatic symptoms—including headache, sleep difficulty, limb pain, and presyncope [13, 28]—which contribute to poor quality of life [29]. FGIDs are inconsistently associated with single parent household, parent with gastrointestinal complaints, mother with neuroticism, or low parental academic attainment [30]. Multiple pediatric studies have demonstrated no association between FGIDs and socioeconomic status (SES) [13, 28, 31, 32].

The morbidity associated with FGIDs is mainly psychosocial—with symptoms interfering with school attendance, performance, relationships, and interactions with family members [33–36]. Quality of life in patients with FGIDs is

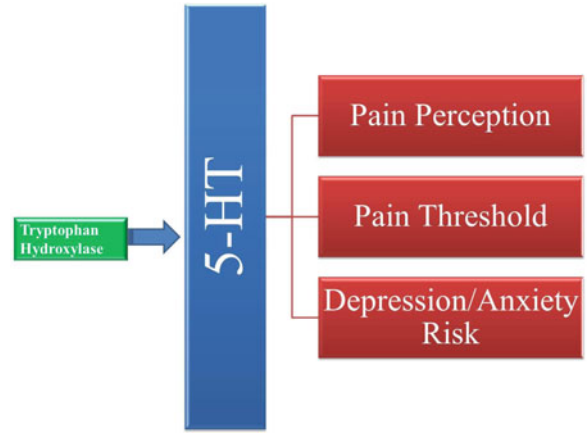

Fig. 5.1 Postulated associations between serotonin (5-HT) and FGIDs

worse than in the general population [37], and similar to that of patients' with organic gastrointestinal disease [38]. Patients with FGIDs also have worse functional impairment and psychiatric symptoms than controls [39–41].

General Etiology and Pathophysiology of FGIDs

Genetic

Several different genetic mutations have been shown to be linked to FGIDs, though causality has not been proven. Thus, mode of inheritance (e.g., autosomal dominant and autosomal recessive) has yet to be uncovered. Genetic polymorphisms in the serotonin transporter promoter, tryptophan hydroxylase, Cathechol-*O*-methyltransferase (COMT) enzyme, IL-10, TNF, and G-protein-coupled receptors have been postulated to be involved in FGIDs.

The neurotransmitter serotonin (5-hydroxytryptamine, 5-HT) has been postulated to control GI motility, secretion, and visceral perception [42, 43]. Studies have noted variations in 5-HT levels [44], and signaling in patients with IBS [45]. The serotonin transporter protein is involved in 5-HT reuptake after it has interacted with its downstream receptor. Thus far, associations between 5-HT transporter promoter variants and IBS have been uncovered but have unclear significance [46], and geographical and gender variation [47–50]. Insights into the pathophysiology behind the 5-HT transporter and IBS have been demonstrated by studies showing that variants can modify the risk for depressive and anxiety episodes in IBS [51], be associated with lower rectal distension pain thresholds in patients with IBS [52], be associated with pain perception itself [53] and differential cerebral blood flow changes in response to colorectal distension in patients with IBS [54]. Furthermore, tryptophan hydroxylase catalyzes the rate-limiting enzyme in the biosynthesis of 5-HT, and Jun et al. [55] noted an association between mutations in the tryptophan hydroxylase gene and diarrhea predominant IBS. These postulated associations are illustrated in Fig. 5.1.

Cathechol-*O*-methyltransferase (COMT) is an enzyme responsible for metabolizing catecholamines. Mutations in the gene encoding for COMT have been found in patients with IBS and are associated with reduced presynaptic dopamine release—which may lead to impaired downregulation of the startle response to threats [56]. The resulting impairment may explain the altered response to pain and resulting hypervigilance in patients with IBS [56].

Mutations in the genes encoding the cytokines IL-10 (decreased in inflammatory states) and TNF (increased in inflammatory states) have been associated with FGIDs, namely IBS [57–59]. Pathophysiologically, these genetic mutations may lead to microscopic patchy inflammation that may not be picked up endoscopically, or that is in areas of the small intestine

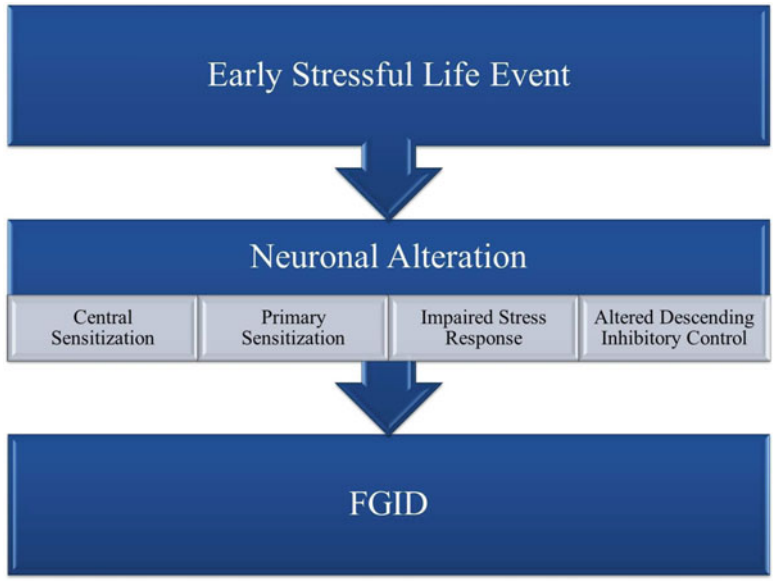

Fig. 5.2 The postulated relationship between early stressful life events and the development of FGIDs

that are unable to be reached by conventional esophagogastroduodenoscopy and colonoscopy.

Other potentially controversial but notable genetic polymorphisms associated with FGIDs include those in G-protein-coupled receptors (GPCRs) [60, 61], cholecystokinin (CCK) [62], and adrenergic receptors [63].

Extrapolating these genetic findings to clinical association studies, it has been noted that 33 % of patients with IBS reported a positive family history of IBS compared to only 5 % of non-IBS control patients [64] and parents of children with FGIDs showed a higher prevalence of FGIDs compared with the parents of children without FGIDs [65]. Many studies have noted high concordance rates for IBS in monozygotic (17–33.3 %) compared to dizygotic twins (8–13.3 %) [66–69], though this notion is somewhat debatable [70]. Environmental factors can confound heritability studies. Finally, the heritability of mental illness may play a role in the development of IBS [69].

Environmental

Environmental factors involved in FGIDs can be categorized into potential early life events, food intolerances, postinfectious phenomena, and small intestinal bacterial overgrowth.

Early Life Events

Early stressful life events may play a role in priming children to develop FGIDs. The mechanisms involved include (1) central (spinal) neuron sensitization, (2) primary (sensory) neuron sensitization, (3) impaired stress response, and (4) altered descending inhibitory control [71] (Fig. 5.2).

Evidence for central neuron sensitization has come from animal studies. It has been demonstrated that repeated neonatal colorectal distension or colonic irritation leads to the development of visceral hypersensitivity [72]. Furthermore, showing that noxious somatic stimulation leads to gut-related visceral hypersensitivity (presumably through impaired visceromotor reflex to colorectal distension) [73] suggests that there is convergence between visceral afferent neurons and somatic neurons in the thoracic—sacral spinal cord. Others have used animal models to show that repetitive exposure to colorectal distension leads to subsequent lower thresholds and higher responses to colorectal distension, as well as increased background neuronal firing in adulthood compared to adult rats that had not been exposed to colonic irritation [74]. These data point to the role of a sensitization mechanism in the development of visceral hypersensitivity.

In addition to mechanical stress, environmental stress also can play a role in visceral hypersensitivity, Coutinho et al. [75] also stressed neonatal rats, but this time by maternal separation for the first 2 weeks of life. Visceral hypersensitivity was deemed present in these rats since their visceromotor reflex to colorectal distension was found to be of higher contractility than those adult rats that were not stressed. Furthermore, these maternally deprived rats have impaired hypothalamic–pituitary–adrenal (HPA) axis responses [76]. These data suggest that stress via activation of the HPA axis augments response to pain.

Descending inhibitory control of pain pathways are immature at birth and can be altered by early life stress. For example, the fact that an opioid receptor antagonist can increase visceral hypersensitivity in rats that are not handled (socially deprived) suggests the possibility of diminished endogenous opioid release in the descending inhibitory pathway as a mechanism for this interaction [75]. This fits with human studies showing that a history of childhood trauma [77] and child abuse [17] have been associated with visceral hypersensitivity (and HPA axis impairment). These psychosocial aspects of a patient's history can provide important treatment targets.

Food Intolerances

Fructose and lactose are specific sugar foods that have been investigated in relation to FGIDs. Thus far, the role for fructose malabsorption in persistent unexplained abdominal pain has been suggested by one small pediatric study [78]. Lactose has been extensively investigated in relation to FGIDs in pediatrics and no causality has been proven [79]. Food allergies themselves have been reported by patients to be a cause of their FGID, but not proven (i.e., by the gold standard double-blind placebo-controlled food challenge) [17]. Gluten sensitivity has been postulated to be involved in IBS [80] though it may be a separate entity itself, and its discussion is beyond the scope of this chapter. Overall, there is no strong data suggesting a role for food intolerances or allergies in the development of FGIDs.

Enteric Infections

Enteric infections may lead to FGIDs in some patients. Acute infectious gastroenteritis is common and occurs roughly 1.4 times per year per person. A subset (7–31 %) of people go on to develop postinfectious FGIDs (PI-FGIDs) [81]. Furthermore, meta-analyses have estimated the relative risk of having IBS 1 year after suffering from an acute gastroenteritis to be increased by approximately sevenfold [82, 83]. Thus far, potential organisms include viruses (rotavirus), bacteria (*Salmonella*, *Shigella*, and *Campylobacter*), and parasites (*Giardia* and Blastocystis) [81]. The caveat behind all of these studies is proving causality and finding the offending organism.

Risk factors for developing PI-FGIDs include younger age of infectious onset, longer illness duration, psychiatric factors (comorbid anxiety and depression, hypochondriasis), psychosocial factors (adverse life events in the preceding 3 months), organism type (higher rates for *Campylobacter* and *Shigella* compared to *Salmonella*), mucosal markers of inflammation, treatment with antibiotics, and possibly female sex [81, 84, 85]. The putative pathogenetic mechanisms of PI-IBS is a transient infectious gastroenteritis that leads to the persistence of a mild inflammatory response (T cells and mast cells) and enterochromaffin cell hyperplasia as well as increased mucosal permeability. Inflammatory cells release cytokines and enterochromaffin cells release serotonin (5-HT). These mediators lead to intestinal motor dysfunction and increased sensory perception [81, 86–88]. Gradual recovery from postinfectious FGIDs typically occurs over years [83].

Along the lines of infectious agents in the gut, the relationship of small intestinal bacterial overgrowth (SIBO) has been investigated in patients with FGIDs and thus far, no specific relationship has been proven [89, 90].

Psychological

Psychopathology is presented in roughly 50 % of patients with FGIDs [91] and has been postulated to play a role in the development and perpetuation of FGIDs. Although there is a notion of

tertiary referral center bias and psychological factors influencing health-seeking behavior [92, 93], even community-based studies have associated psychological dysfunction in patients with FGIDs [91, 94–96]. Most of the literature investigating psychological comorbidities and FGIDs has centered on functional dyspepsia and IBS. In those two disorders, depression, anxiety, and somatization disorders have been shown to be comorbid in pediatric [25, 97, 98] and adult studies [99–104]. Furthermore, the degree of depression and anxiety has been shown to be associated with GI symptom severity [105]. Research has shown catastrophic thinking, even in the absence of depression, negatively influences the perception of pain in FGIDs [106], potentially through increased worrying [107].

Psychopathology (depression, anxiety, and somatization) can lead to and/or be the result of FGID symptoms.

The proposed pathophysiology behind the relationship between psychological disorders and FGIDs revolves around the central nervous system involved in emotional and visceral sensation leading to defective regulation of the HPA axis, autonomic nervous system, and enteric nervous systems. Alternatively, mood and anxiety disorders can predispose the brain to misinterpret or negatively perceive visceral cues. There is a high degree of psychological dysfunction in patients with FGIDs that may affect symptom perception and healthcare-seeking behavior. However, explaining the relationship between psychopathology and functional symptoms to patients in attempts to guide treatment is often difficult, as unfortunately many patients are reluctant to accept the diagnosis of a concomitant psychological disorder. Thus, it is important for patients to hear from the primary care physician that the central processing of pain is an important, treatable risk factor contributing to the severity of their FGID symptoms.

Sleep

Sleep disturbance is associated with FGIDs [108, 109], though their relationship is unclear. Functional symptoms, though rare overnight, may lead to sleep disturbance and vice versa—where increased arousal could lead to chronic stress and altered visceral sensitivity. Nevertheless, it is important to screen for sleep dysfunction in patients with FGIDs as it may guide treatment. A sleep evaluation can be achieved by asking about different components of sleep disturbance such as quality of sleep, restorative nature upon awakening, nighttime awakenings, difficulty falling asleep, and degree of daytime sleepiness or naps. Based on this clinical evaluation, a sleep study may be considered.

Psychosocial

Psychosocial dysfunction does exist in patients with FGIDs, and studies have specifically shown an association between poor quality of life [38], psychosocial distress, emotional instability [110], environmental stressors, and poor coping skills with FGIDs [102, 111–113]. It is thus far unclear whether psychosocial dysfunction induces illness behavior, is caused by the FGID itself, or both situations have an influence [102, 114, 115].

As mentioned earlier, coping skills impact FGID symptom severity, course, and disability. These coping skills are influenced by patients' social network and their own symptom mastery efforts. There are four types of coping styles [116]—engaged, dependent, self-reliant, and avoidant copers. Avoidant copers withdraw from relationships and poorly master symptoms. Symptoms are viewed as serious and patients feel powerless. Dependent copers do little to master symptoms themselves. Unlike avoidant copers, they seek social support, but the comfort derived from support elicited by their helplessness/catastrophizing may reinforce their disability. Unlike the previous two, self-reliant copers are stoic. They attempt to master their symptoms without letting others know. Yet, their stoicism can lead to depression. Lastly, engaged copers have the optimal response—they are the most resilient to symptoms and have an adaptive response.

Parents may play a role in modulating symptom perception and illness behavior in children. In a meta-analysis examining this postulate [117], causality could not be proven. Yet, there is emerging literature noting parental catastrophizing as a

mediator [118] and parental in addition to child therapy in intervention studies [119] also suggests a parental role for symptom perception and report in FGIDs in children.

Overall in the psychosocial realm, there is strong data suggesting that catastrophic thinking, coping strategies, and parental responses to patient symptoms have an etiological role for FGID symptoms and are definite targets for modulation in therapy.

Visceral Hypersensitivity

The aforementioned factors may influence the well-documented association of visceral hyperalgesia in FGIDs [120–122]. The pathophysiology behind this visceral hypersensitivity is multifactorial and may involve the HPA axis, autonomic nervous system, and limbic system. Psychological (namely anxiety and depression), psychosocial (i.e., somatization), genetic, and environmental factors likely modulate these pathways [92, 123]. Moreover, recent adult [124] and pediatric [125] data suggest symptom report, not necessarily visceral hyperalgesia, is heightened in FGIDs. Thus, when guiding therapy, clinicians have to consider the multifactorial nature behind symptom perception and report, and the current modalities available to address dysfunction in these realms.

General Differential Diagnosis for FGIDs

Organic gastrointestinal conditions can be difficult to diagnose and may mimic symptoms of FGIDs. The general categories of these etiologies are detailed in Table 5.1.

How Potentially Comorbid FGIDs Can Affect the Management of Organic Disorders?

The premise behind functional symptoms in intestinal inflammatory (organic) conditions is that gut inflammation can cause functional and

Table 5.1 General categories for differential diagnosis of FGIDs

Intracranial
• Chiari malformation
• Mass
• Pseudotumor cerebri
Infectious
• Enteric parasitic infection—giardiasis and cryptosporidiosis
• *H.* pylori gastritis
• *C. difficile*
• Urinary tract infection
• Otolaryngologic infections
Inflammatory
• Inflammatory bowel disease
• Celiac disease
• Autoimmune enteropathy
• Peptic ulcer disease
• Henoch–Schonlein purpura
Anatomic
• Malrotation with intermittent volvulus
• Intermittent intussusception
• Intestinal stricture
• Intestinal duplications
• Otolaryngologic clefts
Intolerance
• Eosinophilic gastrointestinal disorders
• Lactose intolerance
• Fructose intolerance
• Gluten sensitivity
• Artificial sweeteners
Pancreaticobiliary
• Biliary colic
• Choledocholithiasis
• Cholangitis
• Chronic pancreatitis
Genitourinary
• Urinary tract infection
• Nephrolithiasis
Gynecologic
• Dysmenorrhea
• Endometriosis
• Pelvic inflammatory disease
Dietary
• Vitamin deficiency/toxicity
• Heavy metal toxicity
Psychological
• Anorexia nervosa
• Bulimia
• Somatization

(continued)

Table 5.1 (continued)

• Conversion disorder
• Pain disorder
• Malingering
• Hypochondriasis
• Munchausen's
Miscellaneous
• Acute intermittent porphyria
• Hereditary angioedema

structural central nervous system changes [126] and potential autonomic dysregulation [127]. Additionally, psychological and psychosocial underpinnings in these conditions can affect symptom perception and report. Two organic conditions where comorbid FGIDs have been studied include Celiac disease and Inflammatory Bowel Disease (IBD).

Inflammatory Bowel Disease

Inflammatory bowel disease (IBD) comprises a group of immunodysregulatory disorders, namely Crohn's disease (CD) and Ulcerative Colitis (UC), whose main manifestation is gut inflammation. The peak ages of onset are 15–30 years and 50–70 years, respectively, though the condition can arise at any age [128]. The disease is characterized by variable time periods of flares and remission, and is lifelong without a cure. Treatment consists of immunosuppressive medications and potential surgical resection of diseased bowel and is dictated by the presence of intestinal and extraintestinal symptoms, physical exam, and biochemical markers of inflammation. Symptoms can be nonspecific—including abdominal pain, nausea, and diarrhea—and have disease (e.g., inflammation and postsurgical sequelae) and nondisease-related underpinnings, which potentially complicate management strategies and lead to potential unnecessary escalation of anti-inflammatory medication.

The utility of screening for psychological dysfunction in patients with IBD and potentially functional gastrointestinal symptoms has been demonstrated. There are high rates of FGIDs in patients with inactive IBD in pediatric and adult studies [129, 130]. Psychopathology (anxiety and depression) has been identified as a predictor for an FGID in IBD [130, 131]. Thus, potentially identifying psychosocial dysfunction may help guide more appropriate therapy (i.e., behavioral) for patients with persistent symptoms but otherwise quiescent disease.

Celiac Disease

Celiac disease is a T-cell-mediated gluten-sensitive enteropathy. Patients with this condition have an abnormal immune response leading to small intestinal inflammation to gluten-containing foods such as wheat, rye, and barley. The main sequelae of Celiac disease is malabsorption. Symptoms of Celiac disease can overlap with those of FGIDs—including abdominal pain, diarrhea, bloating, and constipation. This fact can adversely affect the management of Celiac disease and adherence to a gluten-free diet based on somatic gastrointestinal symptoms. The ideal means to assess gluten-free diet adherence and disease remission is serological measurement (which should normalize within 6 months of treatment) and potentially repeat endoscopy (which should demonstrate mucosal lesions have healed within a year of treatment).

Adult studies assessing the association of Celiac disease with irritable bowel syndrome are conflicting [132, 133].

In the most recent pediatric study, the association of Rome III FGIDs with Celiac disease was investigated in a pediatric population [134]. In their study, the authors elucidated symptoms (gastrointestinal and extraintestinal) of patients at the diagnosis of Celiac disease, then after a year following the gluten-free diet. They compared these symptoms to a control population consisting of healthy age and gender matched pediatric patients on a regular diet. After 1 year, all ($N = 82$) of the enrolled patients with Celiac disease were in remission (negative serologies), but 23/82 of these satisfied criteria for a Rome III FGID. The group found functional constipation (18 %) was the most prevalent disorder in pediatric patients with Celiac disease who were in remission. This condition was followed by IBS (7.3 %), FAP (4.8 %), and FD (2 %). 4/23 patients with Celiac disease in remission had an overlap of FGIDs.

Somatic gastrointestinal complaints (rather than objective symptoms such as weight loss) at presentation were associated with an FGID while patients were in remission. Patients with Celiac disease who developed FGIDs had higher rates of anxiety than those who did not develop an FGID.

How to Make a Diagnosis of Fgids?

The diagnosis of FGIDs is made on clinical grounds—mainly based on the Rome III criteria, the absence of red flag symptoms, the presence or absence of psychological and psychosocial pathology, potentially suggestive family history, or previous stressful life events or infections.

Examples of red flag symptoms include but are not limited to unintentional weight loss, linear growth deceleration, evidence of gastrointestinal bleeding, fevers not attributable to an infectious source, vomiting, anemia, or the presence of nocturnal symptoms. All of the studies to date examining the utility of red flags in FGIDs have been done in adults. Notably, Whitehead et al. [135] examined primary (80 %) and GI clinics (20 %) and found 83.3 % of patients with a clinical diagnosis of IBS noted ≥1 red flag, and average was 1.65 per IBS patient. The positive predictive value of red flags for identifying IBD or malabsorption was 7–9 %. Vanner et al. [136] examined only adult GI clinics followed patients prospectively for organic diagnosis change and noted positive predictive value of an FGID being >96–98 % if Rome criteria and red flag elimination were used. These data suggest the exclusion of red flag symptoms in addition to utilization of Rome III criteria is extremely helpful for diagnosing FGIDs, and laboratory testing and expensive invasive diagnostic procedures can be avoided.

The Pediatric FGIDs

Abdominal Pain-Related FGIDs

Pediatric IBS

There are a paucity of studies investigating the prevalence of pediatric IBS, and the diagnostic criteria for adult and pediatric IBS are slightly different in terms of frequency and duration of symptoms. In adult studies, prevalence rates range from 10 to 15 %. In pediatrics, Hyams et al. [25] found 14 % of students in grades 10–12 and 6 % of students in grades 7–9 fit the diagnostic criteria for (Rome II) pediatric IBS. Males and females were affected equally and rates were higher in white students than black students. In North America, the female-to-male ratio for IBS prevalence averages 2:1 in community surveys and closer to 4:1 in referral centers [1].

Pediatric IBS is also associated with significant psychosocial dysfunction [25, 137]. Furthermore, compared to controls, patients with IBS (and functional abdominal pain) on 5 year follow-up have more abdominal pain, somatic pain, functional impairment, and psychiatric dysfunction [39, 138].

Of the abdominal pain-related FGIDs, depending on the study setting, pediatric IBS is either the most common or second most common AP-related FGID [25, 139].

The postulated pathophysiology of IBS is complex and described above. The Rome III diagnostic criteria for pediatric IBS are noted below. The diagnosis of IBS is made clinically and requires physician discretion on the utility of biochemical, imaging, and endoscopic modalities based on the presence or absence of red flag symptoms or other suggestive findings noted above on history. Other conditions to consider in the differential diagnosis for pediatric IBS are noted in Table 5.1.

Pediatric IBS

Diagnostic criteria* Must include **both** of the following:

1. Abdominal discomfort** or pain associated with two or more of the following at least 25 % of the time:
   (a) Improvement with defecation
   (b) Onset associated with a change in frequency of stool
   (c) Onset associated with a change in form (appearance) of stool
2. No evidence of an inflammatory, anatomic, metabolic, or neoplastic process that explains the subject's symptoms

*Criteria fulfilled at least once per week for at least 2 months prior to diagnosis.

**"Discomfort" means an uncomfortable sensation not described as pain.

Functional Dyspepsia

The prevalence of pediatric FD is 5–16 % in the USA [19, 25]. The condition is typically characterized by epigastric pain and reflux with symptoms commonly occur after eating.

The differential diagnosis for FD in pediatrics includes GERD, peptic ulcer disease, Celiac disease, IBD, and drug side effects (NSAIDs, iron, and antibiotics). This differential is much smaller than that of adults, mainly because the incidence of gastric and esophageal cancer is very low. Symptomatic cholelithiasis has not been proven to have an association with dyspepsia. Similarly, although they can be similar in location of symptoms, acute pancreatitis symptoms typically are more severe and have more acute presentation [140]. Specific food intolerances have not been proven to be associated with (functional) dyspepsia [141]. Although *H. pylori* can affect the stomach and duodenum, the incidence and prevalence of *H. pylori* in developed countries such as the USA is very low [142], and although it does have an association with FD more recently [143], there is a dearth of pediatric data.

Aside from the general etiologies of FGIDs described above, the etiologies of FD have not been fully delineated. Delayed gastric compliance, gastric dysmotility, autonomic dysfunction, psychological (anxiety), and psychosocial (stress) factors may play separate or combined roles in symptom pathophysiology [144–148]. Other factors include the interaction of increased acid production and the development of visceral hypersensitivity [149].

The Rome III diagnostic criteria for FD are noted below. As the diagnosis of FD is made clinically, history, examination, and potentially FOBT testing can determine the utility in further workup. Of note, the utility of the lack of red flag symptoms mitigating against endoscopy for dyspeptic symptoms has been demonstrated in a recent study [150].

Functional Dyspepsia

Diagnostic criteria Must include* ***all*** *of the following:*

1. Persistent or recurrent pain or discomfort centered in the upper abdomen (above the umbilicus)
2. Not relieved by defecation or associated with the onset of a change in stool frequency or stool form (i.e., not irritable bowel syndrome)
3. No evidence of an inflammatory, anatomic, metabolic, or neoplastic process that explains the subject's symptoms

*Criteria fulfilled at least once per week for at least 2 months prior to diagnosis.

Functional Abdominal Pain

Chronic or recurrent abdominal pain affects 10–12 % of pediatric patients [145, 151]. A majority of those patients have an abdominal pain-related FGID, while less than 25 % of them have an organic cause for their pain [18, 152].

The prevalence of pediatric abdominal pain-related FGIDs is 0.5–19 % in community-based studies [145, 153, 154]. AP-related FGIDS account for ~7.5 % of GI clinic visits [19, 20]. The variation in these rates is due to the diagnostic criteria used for AP-related FGIDs—Apley's criteria for recurrent abdominal pain (which can include organic disorders [154]), and Rome II versus Rome III (less stringent [21]). A clinic-based study found that FAP was the most common AP-associated FGID [139].

The pathophysiology behind FAP is complex, multifactorial, and outlined in detail above. In general, genetic, postinfectious, environmental (early life events and psychosocial stressors), alterations in sensory processing, and intestinal dysmotility all play a role to different extents in pediatric patients with FAP.

The differential diagnosis for FAP is vast and noted in Table 5.1.

The Rome III diagnostic criteria for the two subtypes of FAP are listed below. The diagnosis of this disorder is made clinically based on history and physical exam with careful attention paid to signs/symptoms of the above conditions on the differential diagnosis, psychological and psychosocial dysfunction, and red flag symptoms.

Frequency, severity, location, and timing of pain do not distinguish organic from FAP [155]. In the absence of suggestive symptoms/signs of an organic disorder, there is no need for further diagnostic workup. Important factors to note on history include distractibility, overreporting of symptoms, minimizing psychological/psychosocial contributors to symptoms, requesting further diagnostic studies, and seeking of frequent healthcare [156].

Childhood Functional Abdominal Pain

Diagnostic criteria Must include* ***all*** *of the following:*

1. Episodic or continuous abdominal pain
2. Insufficient criteria for other FGIDs
3. No evidence of an inflammatory, anatomic, metabolic, or neoplastic process that explains the subject's symptoms

*Criteria fulfilled at least once per week for at least 2 months prior to diagnosis.

Childhood Functional Abdominal Pain Syndrome

Diagnostic criteria Must satisfy criteria for childhood functional abdominal pain and have at least 25 % of the time* ***one or more*** *of the following:*

1. Some loss of daily functioning
2. Additional somatic symptoms such as headache, limb pain, or difficulty sleeping

*Criteria fulfilled at least once per week for at least 2 months prior to diagnosis.

Abdominal Migraine

Abdominal migraine is a variant of migraine headaches that affects 1–4 % of pediatric patients. The condition has a female predominance and usually occurs between the ages of 7 and 12 years. The pathophysiology behind abdominal migraines is unclear but may involve the interplay between food allergy, neurotransmitter metabolism, and abnormal gut permeability. These factors may be aggravated by stress [157].

The differential diagnosis for abdominal migraine is vast and includes causes for an acute abdomen (appendicitis, peritonitis, and intestinal perforation), acute abdominal pain (intestinal inflammation, acute pancreatitis, biliary colic, choledocholithiasis, cholangitis, nephrolithiasis, and ureteropelvic junction obstruction), and chronic abdominal pain (listed above). History and physical exam is aimed to rule out the above conditions, as the diagnosis of abdominal migraine is a diagnosis of exclusion.

Abdominal Migraine

Diagnostic criteria Must include* ***all*** *of the following:*

1. Paroxysmal episodes of intense, acute periumbilical pain that lasts for hour or more
2. Intervening periods of usual health lasting weeks to months
3. The pain interferes with normal activities
4. The pain is associated with two of the following:
   (a) Anorexia
   (b) Nausea
   (c) Vomiting
   (d) Headache
   (e) Photophobia
   (f) Pallor

No evidence of an inflammatory, anatomic, metabolic, or neoplastic process considered that explains the subject's symptoms.

*Criteria fulfilled two or more times in the preceding 12 months.

Constipation and Incontinence

Functional Constipation

Functional constipation accounts for 3 % of general pediatric outpatient visits and 10–25 % of pediatric gastroenterology clinic visits [158, 159]. Functional constipation has a peak incidence in ages 0–4 years, while one of its major sequelae, encopresis, peaks between the ages of 4 and 7 year [158–160]. Although the rates of pediatric constipation continuing into adulthood are unknown [161], age of presentation may affect prognosis. For example, it has been shown that patients who present with functional constipation at a younger age are less likely to have symptoms several years later [162, 163]. Socioeconomic status, family size, parental age, and ordinal position in the family do not affect the incidence of functional constipation.

Table 5.2 Etiology of functional constipation and encopresis

Painful/frightening evacuation	Stress	Behavioral
	– Change in routine	– Oppositional
Toilet training	Fear of public bathrooms	Sexual abuse
Intercurrent illness with associated dysmotility	Too Busy	Dietary
	– Playing	– Dehydration
	– Busy schedule	– Malnutrition
		– Low fiber

The pathophysiology of constipation and potentially resulting encopresis is based on dysfunctional defecation. In normal defecation, stool descends from the sigmoid colon into the rectum and puts pressure on the rectal wall, thereby activating rectal wall stretch receptors. These stretch receptors transmit signals to the S2–S4 defecation center and signal the urge to defecate, at which point the internal anal sphincter is reflexively relaxed, and the external anal sphincter and puborectalis muscle are voluntarily contracted. When it is convenient to defecate, the body voluntarily relaxes the external anal sphincter and contracts the abdominal musculature. If defecation is inconvenient, there is voluntary contraction of the external anal sphincter and puborectalis muscle, and the rectum accommodates the retained stool. If it is chronically either inconvenient for a child to stool or the child does not want to stool for another reason, repeated denial of evacuation leads to rectal stretching and progressive rectal dilation. The dilated rectum has decreased sensory and motor function, and thereby an increased threshold for defecation urge. The longer the stool stays in the rectum, the more water is removed from it, and the less convenient/more painful it is to defecate. Once this hardened fecal mass is large enough, soft stool can leak around it, leading to involuntary fecal soiling. The constantly filled rectum also leads to overflow soiling whenever the child tries to pass gas or is excited. The resulting behaviors of stool withholding can include stiff legs that are often crossed, or bent-over posturing in a corner or behind a piece of furniture. They often appear dance-like and can even be confused with a seizure [164].

Table 5.3 Differential diagnosis for functional constipation

Anatomic
Anal stenosis
Anteriorly displaced anus
Pelvic mass
Metabolic
Hypothyroidism
Electrolyte abnormalities
Hypokalemia
Hypercalcemia
Inflammatory
Celiac disease
Neuromuscular
Spina bifida
Hirschsprung's disease
Intestinal neuronal dysplasia
Visceral myopathy
Prune belly syndrome
Ingestions
Heavy metal
Narcotics
Anticholinergics

The etiology of the initial withholding behavior that can lead to encopresis is listed in Table 5.2 and could be multifactorial.

The differential diagnosis of functional constipation can be divided into broad categories—anatomic, metabolic, neuromuscular, or medication side effects. These are detailed in Table 5.3.

The Rome III and DSM-IV diagnostic criteria for functional constipation, encopresis, and nonretentive fecal incontinence are listed below. History and physical exam dictates the workup for these diagnoses. Specifically, as the diagnosis of constipation is clinical, history should be based on the above differential diagnosis. The clinician should note stooling frequency, caliber,

and associated symptoms such as straining, dyschezia, blood in stool, and withholding behavior (which can mimic straining). Fecal soiling should be asked about, with specific questioning regarding involuntary versus voluntary. Of note, in addition to examining the neck (thyromegaly), abdomen (fecal masses), back (sacral dimples/tufts), and doing a complete neuromuscular exam, the clinician should perform a rectal exam specifically looking at rectal tone, presence/absence of anal wink, and the caliber and size of stool in the rectal vault.

Functional Constipation

Diagnostic criteria Must include **two or more** of the following in a child with a developmental age of at least 4 years with insufficient criteria for diagnosis of IBS*:

1. Two or fewer defecations in the toilet per week
2. At least one episode of fecal incontinence per week
3. History of retentive posturing or excessive volitional stool retention
4. History of painful or hard bowel movements
5. Presence of a large fecal mass in the rectum
6. History of large diameter stools which may obstruct the toilet

* Criteria fulfilled at least once per week for at least 2 months prior to diagnosis.

Functional Fecal Incontinence

*Diagnostic criteria**

1. Recurrent uncontrolled passage of fecal material in an individual with a developmental age of at least 4 years and *one or more* of the following:
2. Abnormal functioning of normally innervated and structurally intact muscles
3. Minor abnormalities of sphincter structure and/or innervation
4. Normal or disordered bowel habits (i.e., fecal retention or diarrhea)
5. Psychological causes

AND

Exclusion of *all* the following:

(a) Abnormal innervation caused by lesion(s) within the brain (e.g., dementia), spinal cord, or sacral nerve roots, or mixed lesions (e.g., multiple sclerosis), or as part of a generalized peripheral or autonomic neuropathy (e.g., due to diabetes)
(b) Anal sphincter abnormalities associated with a multisystem disease (e.g., scleroderma)
(c) Structural or neurogenic abnormalities believed to be the major or primary cause of fecal incontinence

* Criteria fulfilled for the last 3 months

Nonretentive Fecal Incontinence

Diagnostic criteria Must include **all** of the following in a child with a developmental age at least 4 years:*

1. Defecation into places inappropriate to the social context at least once per month
2. No evidence of an inflammatory, anatomic, metabolic, or neoplastic process that explains the subject's symptoms
3. No evidence of fecal retention

*Criteria fulfilled for at least 2 months prior to diagnosis.

Upper Gastrointestinal Disorders

Functional Dysphagia

Functional dysphagia is a rare FGID, though its exact prevalence has not yet been accurately assessed. The hallmark of the disorder is the sensation of abnormal bolus passing through the esophagus.

The three main pathophysiologic features of functional dysphagia include potential peristaltic dysfunction (rapid transit followed by poor clearance) [165], poor esophageal emptying alone [166], and visceral hypersensitivity of the esophagus [167]. Psychological underpinnings may be modifiers of these features [168].

The diagnosis of functional dysphagia is noted below. The differential diagnosis includes esophageal structural obstruction (stricture, external compression, and foreign body ingestion), esophageal inflammation (GERD and Eosinophilic esophagitis), motility disorder (Achalasia, nutcracker esophagus, less likely but possibly diffuse esophageal spasm). Given the wide range of conditions on the differential diagnosis for functional dysphagia, the diagnosis should not be

purely made on a solely clinical basis. If a patient fails an empiric PPI trial, radiologic tests, endoscopies, and if history suggestive, esophageal manometry, should be undertaken to rule out the aforementioned conditions on the differential diagnosis.

Functional Dysphagia

Diagnostic criteria Must include* ***all*** *of the following:*

1. Sense of solid and/or liquid foods sticking, lodging, or passing abnormally through the esophagus
2. Absence of evidence that gastroesophageal reflux is the cause of the symptom
3. Absence of histopathology-based esophageal motility disorders

* Criteria fulfilled for the last 3 months with symptom onset at least 6 months prior to diagnosis

Globus

Globus is a persistent feeling of a lump in the throat. The symptom is usually episodic and improves with eating.

The symptom of globus reported in up to 50 % of people in the USA, uncommon in patients less than 20 years old [169], and equally prevalent in males and females [170].

The pathophysiology behind globus is thus far unclear. Postulates include air entrapment in the proximal esophagus caused by the urge to swallow and increased swallow frequency [171] and/or esophageal hypersensitivity [172]. Diagnosable psychological disorders have not been proven to be etiological in globus. However, psychosocial factors that may lead to this disorder in terms of symptoms or exacerbations include life stress [173] and levels of high emotional intensity [174].

The differential diagnosis for globus includes: Cricopharyngeal achalasia, GERD, esophageal motility disorders, external compression (i.e., goiter), or Chiari malformation.

The diagnostic criteria for globus are noted below. It is important to note the absence of dysphagia and odynophagia for this diagnosis. In addition to clinical history and physical exam (including oropharyngeal exam), consideration for laryngoscopy, swallow study, esophagram, head MRI, or even therapeutic trial of a PPI is worth noting. Alarm symptoms that may lead away from the diagnosis of globus but may prompt even further investigation are: dysphagia, odynophagia, pain, weight loss, and hoarseness.

Diagnostic Criteria* for Globus

Must include *all* of the following:

1. Persistent or intermittent, nonpainful sensation of a lump or foreign body in the throat
2. Occurrence of the sensation between meals
3. Absence of dysphagia or odynophagia
4. Absence of evidence that gastroesophageal reflux is the cause of the symptom
5. Absence of histopathology-based esophageal motility disorders

**Criteria fulfilled for the last 3 months with symptom onset at least 6 months before diagnosis.*

Chronic Idiopathic Nausea

The epidemiology of chronic idiopathic nausea (CIN) is unknown [175].

The pathophysiology of CIN is not fully understood, though causative factors include visceral hypersensitivity [176], altered gut flora [177], and dysmotility. There is no association between CIN and psychological disorders, but stress and psychosocial factors have been thought to play a role [178].

GERD, gastroparesis, metabolic abnormalities (hypokalemia, hypercalcemia, and uremia), rumination syndrome, intracranial mass, intestinal stricture, and esophageal webs are all conditions that exist on the differential diagnosis for CIN.

In addition to a history and physical that attempts to rule out other conditions on the differential diagnosis, diagnostic testing to rule out CIN is essential. Clinicians should consider the following tests: neuroimaging, electrolytes, endoscopy, and potentially small bowel imaging. Other modalities that may be helpful diagnostic tools include an empiric PPI trial or gastric emptying scan.

Chronic Idiopathic Nausea

Diagnostic criteria Must include all of the following:*

1. Bothersome nausea occurring at least several times per week
2. Not usually associated with vomiting
3. Absence of abnormalities at upper endoscopy or metabolic disease that explains the nausea

*Criteria fulfilled for the last 3 months with symptom onset at least 6 months prior to diagnosis.

Aerophagia

Aerophagia is characterized by excessive (too frequent or too large quantities) swallowing of air. This leads to significant gaseous distension of the intestinal tract.

Aerophagia is rare in healthy children—with rates of 1.3 % of pediatric patients in a pediatric gastroenterology clinic [20]. It is more common in patients with mental retardation [179] and after fundoplication [180].

The differential diagnosis for aerophagia includes GERD, anatomic obstructive disorders, intestinal inflammatory disorders, and carbohydrate malabsorption. The Rome III Diagnostic Criteria for aerophagia are listed below. The diagnosis of aerophagia lies in the fact that patients have increasing abdominal distension during the day that resolves overnight. Associated symptoms include excessive eructation, vomiting, abdominal pain, and flatulence. If there are no red flags on history or physical exam suggestive of the aforementioned disorders on the differential diagnosis, the diagnosis of aerophagia can be made on a clinical level.

Aerophagia

Diagnostic criteria Must include **at least two** of the following:*

1. Air swallowing
2. Abdominal distention due to intraluminal air
3. Repetitive belching and/or increased flatus

*Criteria fulfilled at least once per week for at least 2 months prior to diagnosis.

Disorders of Infants and Toddlers

Functional Diarrhea

Functional diarrhea in pediatrics is a self-limited disorder characterized by frequent loose stools that often contain undigested food in individuals who are otherwise growing and well-appearing. Although dietary factors have been proposed to be implicated, the pathophysiology is unknown.

The differential diagnosis for functional diarrhea is vast and is listed in Table 5.1. The Rome III criteria for Functional Diarrhea are below. The diagnosis of functional diarrhea rests on history and physical examination. The clinician should ask about recent travels, fevers, antibiotic use, laxatives, excessive fruit juice or artificial sweeteners, diet changes, constitutional symptoms, or signs of blood in the stool. History will often note stools containing visible undigested food and/or mucus. Physical exam mainly examines the growth chart for failure to thrive and signs of malnutrition on exam.

Pediatric Functional Diarrhea

*Diagnostic criteria Must include **all** of the following:*

1. Daily painless, recurrent passage of three or more large, unformed stools
2. Symptoms that last more than 4 weeks
3. Onset of symptoms that begins between 6 and 36 months of age
4. Passage of stools that occurs during waking hours
5. There is no failure-to-thrive if caloric intake is adequate

Rumination Syndrome

Rumination syndrome is a behavioral disorder consisting of the effortless regurgitation of recently ingested food followed by either spitting up or rechewing and reswallowing. Patients with rumination syndrome do not have any abdominal pain, nausea, or heartburn when the act occurs. Although most commonly seen in infants (see Rome III criteria below) and developmentally disabled individuals, rumination syndrome can occur in healthy children, too. Rumination syndrome usually occurs immediately after a meal, and patients usually stop regurgitating when the gastric contents become acidic [181]. Associated comorbidities include depression, post-traumatic stress disorder, and somatoform disorders.

The differential diagnosis for rumination syndrome includes GERD, gastroparesis, bulimia, and obstructive intestinal pathology. The Rome III criteria for rumination syndrome are noted below. Careful history will elicit the main symptoms being effortless, a lack of gastrointestinal complaints, and potential psychopathology. Additionally, impedance monitoring combined with manometry may be helpful [182].

Infant Rumination Syndrome

Diagnostic criteria Must include ***all*** *of the following for at least 3 months:*

1. Repetitive contractions of the abdominal muscles, diaphragm, and tongue
2. Regurgitation of gastric content into the mouth, which is either expectorated or rechewed and reswallowed
3. Three or more of the following:
   (a) Onset between 3 and 8 months
   (b) Does not respond to management for gastroesophageal reflux disease, or to anticholinergic drugs, hand restraints, formula changes, and gavage or gastrostomy feedings
   (c) Unaccompanied by signs of nausea or distress
   (d) Does not occur during sleep and when the infant is interacting with individuals in the environment

Adolescent Rumination Syndrome

Diagnostic criteria Must include* ***all*** *of the following:*

1. Repeated painless regurgitation and rechewing or expulsion of food that
   (a) begin soon after ingestion of a meal
   (b) do not occur during sleep
   (c) do not respond to standard treatment for gastroesophageal reflux
2. No retching
3. No evidence of an inflammatory, anatomic, metabolic, or neoplastic process that explains the subject's symptoms

*Criteria fulfilled for the last 3 months with symptom onset at least 6 months prior to diagnosis.

Case Studies

Case 1

Kyleigh is a 12-year-old premenarchal female who presents to the GI clinic complaining of moderate severity, constant, and periumbilical abdominal pain since starting middle school several months ago. The pain is never nocturnal. Kyleigh denies weight loss, fevers, fatigue, oral ulcers, nausea, vomiting, regurgitation, heartburn, constipation, blood in stool, rashes, difficulty sleeping, headache, or limb pain. She eats a well-balanced diet. Kyleigh worries about eating at times but has not found any one particular food that seems to bother her. On social history, Kyleigh mentions although she has not missed any school, she does not like middle school, and upon further investigation notes her stomach pain decreases over the weekends.

On exam, the patient's height, weight, and body mass index are in the 50th percentile and tracking appropriately over time. She is well-appearing, and in no distress. She has distractible diffuse mild abdominal tenderness on exam. There is no scleral icterus, conjunctival pallor, perianal abnormalities (skin tags and fissures), heme-positive stool in the rectal vault, or digital clubbing.

Based on the patient's benign physical exam, distractible abdominal tenderness, situational nature of her abdominal pain, and lack of any red flags symptoms, you make the diagnosis of Rome III Childhood FAP. The history and physical exam mitigate against biochemical or imaging workup at this point.

Questions

1 Which of the following could have led to the patient's FGID?
   (a) Lactose intolerance
   (b) Small intestinal bacterial overgrowth
   (c) Psychological dysfunction
   (d) Previous stressful life event
   (e) (c) and (d)

2 Which of the following differentiates Rome III Childhood FAP *Syndrome* from Rome III Childhood FAP?
   (a) Continuous versus episodic abdominal pain
   (b) The loss of daily functioning
   (c) The presence of somatic symptoms (headache, limb pain, and sleep dysfunction)
   (d) None of the above
   (e) (b) and (c)
3 Which of the following environmental factors have been associated with FGIDs?
   (a) Food allergies
   (b) Sleep dysfunction
   (c) Low socioeconomic status
   (d) Enteric infection
   (e) (b) and (d)

Answers

1. (e): Psychopathology is presented in roughly 50 % of patients with FGIDs [91]. Notably, depression, anxiety, and somatization disorders have been shown to be comorbid in pediatric studies [25, 97]. The proposed pathophysiology behind the relationship between psychological disorders and FGIDs revolves around the central nervous system involved in emotional and visceral sensation leading to defective regulation of the HPA axis, autonomic nervous system, and enteric nervous systems. Early stressful life events may play a role in priming children to develop FGIDs. The mechanisms involved include (1) central (spinal) neuron sensitization, (2) primary (sensory) neuron sensitization, (3) impaired stress response, and (4) altered descending inhibitory control [71]. The relationship of small intestinal bacterial overgrowth with pediatric FGIDs has been speculated, but thus far there has been no proven association [89]. Lactose has been extensively investigated in relation to FGIDs in pediatrics and no causality has been proven [79].
2. (e): The presence of somatic symptoms (headache, limb pain, and sleep dysfunction) and loss of daily functioning differentiates Rome III Childhood FAP Syndrome from Rome III Childhood FAP.
3. (e): Sleep disturbance is associated with FGIDs [108, 109], though their relationship is unclear. Functional symptoms, though rare overnight, may lead to sleep disturbance and vice versa—where increased arousal could lead to chronic stress and altered visceral sensitivity. Meta-analyses have estimated the relative risk of having IBS 1 year after suffering from an acute gastroenteritis to be increased by approximately sevenfold [82, 83]. The caveat behind all of these studies is proving causality and finding the offending organism. Thus far, potential organisms include viruses (rotavirus), bacteria (*Salmonella*, *Shigella*, and *Campylobacter*), and parasites (*Giardia* and *Blastocystis*) [81]. Risk factors for developing PI-FGIDs include younger age of infectious onset, longer illness duration, psychiatric factors (comorbid anxiety and depression, hypochondriasis), psychosocial factors (adverse life events in the preceding 3 months), organism type (higher rates for *Campylobacter* and *Shigella* compared to *Salmonella*), mucosal markers of inflammation, treatment with antibiotics, and possibly female sex [81, 84, 85]. Food allergies themselves have been reported by patients to be a cause of their FGID, but not proven (i.e., by the gold standard double-blind placebo-controlled food challenge) [17]. Socioeconomic status thus far has not been found to have bearing on the rates of FGIDs.

Case 2

Michael is a 17-year-old male who presents to the GI clinic for abdominal pain and weight loss. The abdominal pain insidiously started 3 months ago without inciting change in diet, social setting, or infection. It is periumbilical, cramping, 6/10 in severity, and is associated with early morning awakening (not due to pain). Since the pain emerged, he has noticed a decrease in his appetite and has lost almost 10 lb over 3 months. Michael does not offer any information regarding stress in his life and his parents appear offended when this is questioned during the assessment.

Michael is fatigued, but denies fevers, oral ulcers, nausea, vomiting, regurgitation, heartburn, constipation, blood in stool, or rashes. On review of systems, he notes difficulty concentrating and no enjoyment in his typically fun activities.

On exam, the patient's weight is in the 25th percentile, height in the 50th percentile, and body mass index in the 20th percentile. He has a flat affect but is otherwise well-appearing. He has mild nondistractible periumbilical abdominal tenderness. There is no scleral icterus, conjunctival pallor, perianal abnormalities (skin tags and fissures), heme-positive stool in the rectal vault, digital clubbing, or rashes.

His biochemical workup shows normal hemoglobin, erythrocyte sedimentation rate, C-reactive protein, albumin, liver enzymes, and urinalysis, and negative Celiac serologies. Due to continued concerns for inflammatory bowel disease or Celiac disease, he has a subsequent endoscopy, colonoscopy, and capsule endoscopy—all of which are normal.

Based on the negative workup above and concerns for depression you make a psychology referral who confirms the diagnosis based on further history.

Questions

1. Which of the following psychopathology has been shown to be associated with FGIDs?
   (a) Anxiety
   (b) Depression
   (c) Somatization disorders
   (d) Poor coping skills
   (e) All of the above
2. Which of the following play roles in modulating symptom perception in patients with FGIDs?
   (a) Catastrophic thinking
   (b) Parental response to symptoms
   (c) Transitioning from an avoidant to a dependent coping style
   (d) (a) and (c)
   (e) (a) and (b)
3. Which of the following relationships between psychopathology and FGIDs are true?
   (a) The degree of depression and anxiety have been shown to be associated with the degree of FGID symptom severity
   (b) Psychopathology causes FGIDs
   (c) Both psychopathology and FGIDs can be associated with weight loss
   (d) Both psychopathology and FGIDs can be associated with poor appetite
   (e) (a), (c), and (d)

Answers

1. (e): Anxiety, depression, somatization disorders, and poor coping skills have all been shown to be associated with FGIDs [25, 97, 116].
2. (e): Catastrophic thinking negatively influences the perception of pain in FGIDs [106], potentially through increased worrying [107]. Additionally, parental behavior and their response to their child's symptoms may negatively affect symptom perception and report on behalf of pediatric patients [118, 119] noting parental catastrophizing as a mediator [118]. Parental in addition to child therapy intervention studies [119] also suggests a parental role for symptom perception and report in FGIDs in children. Coping skills impact FGID symptom severity, course, and disability. There are four types of coping styles—engaged, dependent, self-reliant, and avoidant copers. Avoidant copers withdraw from relationships and poorly master symptoms. Symptoms are viewed as serious and patients feel powerless. Dependent copers do little to master symptoms themselves. Unlike avoidant copers, they seek social support, but the comfort derived from support elicited by their helplessness/catastrophizing may reinforce their disability. Neither of these coping styles are adaptive and effective at dealing with functional symptoms [116].
3. (e): There are several overlapping types of psychopathology seen in FGIDs—(1) the characteristics of the psychiatric illness itself, (2) the psychological sequelae of the FGID itself, and (3) psychological factors leading the individual to consult a physician. The relationship between psychological dysfunction and FGIDs is likely bidirectional

and revolves around the central nervous system involved in emotional and visceral sensation leading to defective regulation of the HPA axis, autonomic nervous system, and enteric nervous systems. The degree of depression and anxiety has been shown to be associated with GI symptom severity [105]. Due to potential effects on appetite (and potential fear of postprandial gastrointestinal symptoms or nausea itself), both psychopathology and, albeit, more rarely, FGIDs can lead to weight loss. However, psychopathology does not cause an FGID.

Case 3

Donna is a 16-year-old female who presents to her primary care physician with suspected anorexia nervosa. After several life events (starting home schooling to allow her to focus on being a competitive ballerina, parents going through a divorce), she has lost over 20 lb in the previous year and who has a body mass index less than the 3rd percentile. She notes excellent appetite and oral intake and is content with her current body habitus since she feels it facilitates her to be a better dancer. She denies any bingeing or purging behaviors.

She attributes mild fatigue to her dance practicing. She denies fevers, oral ulcers, nausea, vomiting, abdominal pain, diarrhea, blood in stool, and rashes. On review of systems, she is premenarchal.

On exam, Donna is pale and ill-appearing. Her interactions are appropriate for age. Her abdominal exam notes a fullness in the right lower quadrant, hyperactive bowel sounds, but is otherwise nontender. Her perianal exam demonstrates large skin tags at the 3 o'clock and 10 o'clock positions, with heme-positive soft stool in the rectal vault. She has 1+ digital clubbing.

You note iron-deficiency anemia, hypoalbuminemia, elevated inflammatory markers, and normal white blood cell count, lactate dehydrogenase, and uric acid. Subsequent GI-referral confirms Crohn's disease of the small and large intestine, with a fistula between the duodenum and ascending colon.

Questions

1. Which of the following are NOT historical red flags?
   (a) Unintentional weight loss
   (b) Fevers unexplained by intercurrent illness
   (c) Bilious emesis
   (d) Skin tag at the 6 o'clock position
   (e) Skin tag at the 9 o'clock position
2. Which of the following can be associated with both an eating disorder and inflammatory bowel disease?
   (a) Weight loss
   (b) Anorexia
   (c) Fatigue
   (d) Anemia
   (e) All of the above
3. Which of the following differentiates an eating disorder from a disorder where there is either intestinal malabsorption or a need for increased nutritional requirements?
   (a) Optimum oral intake in the absence of binging or purging behavior
   (b) Primary amenorrhea
   (c) Iron-deficiency anemia
   (d) Fatigue
   (e) None of the above

Answers

1. (d): A skin tag at the 6 o'clock position can be associated with functional constipation, as it is a dependent area when stooling exits from the anus [183]. Unintentional weight loss, fevers unexplained by intercurrent illness, and bilious emesis can all be attributed to organic disorders—inflammatory, oncologic, and potentially obstructive (i.e., intestinal malrotation).
2. (e): Weight loss, anorexia, fatigue (secondary to systemic effects of malnourishment and disease activity), and anemia (vitamin deficiencies secondary to restriction or malabsorption) can be due to an eating disorder or inflammatory bowel disease.

3. (a): Optimum oral intake in the absence of binging or purging behavior differentiates an eating disorder from a disorder where there is either intestinal malabsorption or a need for increased nutritional requirements. Primary amenorrhea can occur secondarily to physiological stress on the body from any of the aforementioned disorders, iron-deficiency anemia can be due to nutritional restriction or the microscopic blood loss, and fatigue can be secondary to malnourishment or systemic inflammation.

Treatment

Many medication and psychological treatment modalities have been studied for FGIDs, and the data on these interventions within the realm of pediatric FGIDs is emerging. Thus far, the strongest treatment data lies in the behavioral treatment realm—where Cognitive–Behavioral Therapy (CBT) for both patient and their families (Chap. 19) and hypnotherapy have been proven to be effective (Chap. 21) [119, 184–187].

Conclusions

As can be seen, FGIDs are common conditions with multiple overlapping etiological underpinnings. Their association with organic gastrointestinal disorders can complicate management of those disorders. As clinical diagnostic criteria have been set forth by the Rome Foundation, a clinical diagnosis is stressed based on history and physical examination, with minimal (or no) diagnostic testing necessary, at least not as a first step. Careful attention to comorbid psychological and psychosocial factors can also increase the likelihood of more appropriate targeted treatment of these conditions while avoiding expensive and unnecessary medical testing. It also appears that many of the precipitating and predisposing factors to develop FGIDs occur during childhood, including in infancy, even if the diagnosis comes much later. Thus, it is important to complete a comprehensive assessment of factors from childhood.

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Functional Symptoms in Nephrology: Keeping It In and Letting It Out

6

Thomas R. Welch

Abstract

Lower urinary tract complaints, generally described by the term "voiding dysfunction," are common in children. While often behavioral and responsive to a number of straightforward interventions, they occasionally signal serious underlying kidney disease. Most children with voiding dysfunction can be diagnosed with a careful history and physical examination and some very minimal further testing. A first morning urinalysis is important for excluding a concentrating defect or proteinuria, either of which could indicate an underlying nephropathy. If there is suspicion of an anatomic abnormality, a renal ultrasound, including views of the bladder before and after voiding, is generally sufficient. Frequent voiding during the day ("functional polyuria") is often self-limited, although self-hypnosis may be used in difficult cases. Nocturnal enuresis ("bedwetting") is ultimately self-limited but may be improved by some techniques such as waking or the use of enuresis alarms. The dysfunctional elimination syndrome often includes both daytime urinary symptoms and constipation. Dietary efforts at controlling constipation, along with timed voiding, often bring about dramatic improvement.

Keywords

Enuresis • Frequent urination • Bedwetting • Voiding dysfunction

Background

The human kidneys have an exquisitely complex mechanism for regulating solute and water balance, the ultimate result of which is the production of urine. The lower urinary tract, consisting of bladder, bladder neck, and urethra, has nothing to do with solute or water balance. It does, however, have the vital function of storing urine as it is continuously produced, and emptying it regularly in a socially appropriate fashion. Disturbances in this storage and emptying function are generally considered "bladder dysfunction" and can cause serious disability. Some causes of bladder dysfunction are clearly anatomic: obstruction of the

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bladder outlet (e.g., posterior urethral valves) and disturbed innervation of the lower urinary tract (e.g., as a result of spina bifida). These should be obvious from history and physical examination and are beyond the scope of this chapter. Beyond these, however, most bladder dysfunction seen in primary care practice is functional and often responsive to simple behavioral intervention. On the other hand, these functional disorders may be confused, or may coexist, with serious anatomic disorders. Distinguishing between these is doubly important. Missing the anatomic basis of a symptom, and treating it as functional, can result in serious deterioration of kidney function. Alternatively, pursuing anatomic explanations for functional complaints can lead to much diagnostic and interventional mischief.

We consider three common urinary tract complaints in children: frequent urination, nocturnal enuresis, and dysfunctional elimination syndrome (DES).

Making the Diagnosis of Functional Urinary Tract Disorders

"Peeing Too Much": Functional Polyuria

Beyond the age of 5 years, it is very unusual for a child to void more than seven times during the day; indeed, the frequency is usually less. Functional polyuria is generally defined as a urine frequency of hourly or more, with void volumes less than half of bladder capacity [1].

The most important differential in a child who is being assessed for frequent urination is whether the problem is actually an *increased volume of urine* rather than increased frequency of urination. There are several clinical clues to aid in this distinction. Children with an increased volume of urine will have urinary frequency at night as well as during the day. They also are likely to drink excess fluid to replace increased urinary losses. Their increased urine frequency will not be affected by environment. Children with functional polyuria, in contrast, may display this behavior only in specific settings (e.g., school).

Acute increases in urine volume may result from the onset of diabetes mellitus or a primary renal disorder associated with a concentrating defect. Thus, the most useful laboratory test to aid in this distinction is a simple urinalysis on a first morning urine. The absence of glucose, protein, and leukocytes makes diabetes mellitus, chronic nephropathy, and urinary tract infection most unlikely. A urine specific gravity of 1.015 or above (assuming no fluid intake during the evening) rules out a primary disorder of urine concentration.

Bedwetting: Primary Nocturnal Enuresis

Children who empty their bladders during sleep, but have bladder control during the waking hours, are said to have primary nocturnal enuresis. The International Children's Continence Society (ICCS) has endorsed the term "enuresis" alone for this phenomenon.

Achieving nighttime dryness is largely a function of central nervous system maturation and is not under voluntary control. A variety of factors, including the age at which parents were dry at night, determine the age at which a child reaches this milestone. Several studies have suggested a rate of spontaneous resolution of enuresis of 15 % per year, making the assessment of any intervention difficult. In most studies, boys are later to achieve nighttime dryness. At 10 years of age, as many as 5 % of boys may still have episodic bedwetting [2].

Primary nocturnal enuresis should generally be diagnosable by a good history and physical examination. There are rarely any diagnostic studies needed in the child who has never achieved nighttime dryness, but who is otherwise normal with no daytime urinary symptoms. Since daytime frequency may rarely be unrecognized in a child with a large bladder capacity, a first morning urinalysis is a reasonable test, as it is for functional polyuria.

Daytime Voiding Symptoms: Dysfunctional Elimination Syndrome

Recognition of the DES may have done more to protect children from potentially harmful diagnostic and therapeutic interventions (including

surgery) than any other development in pediatric urology and nephrology.

Children with DES most often come to attention because of daytime symptoms. These include recurrent urinary tract infection, daytime dribbling of urine, and urgency. Although the family and child may not recognize it initially, DES is very frequently accompanied by stool retention, constipation, and occasionally fecal incontinence [3].

Although studies of the pathophysiology of DES are conflicted, its initiation may be a learned behavior. Contraction of the muscles of the pelvic floor and the external urinary sphincter during bladder contraction result in incomplete bladder emptying. Urinary stasis may predispose to urinary tract infection, and high bladder pressures during voiding may ultimately result in "anatomic" problems such as vesicoureteral reflux. Efforts at urinary withholding may result in behavioral mannerisms such as leg crossing or "Vincent's curtsy." If urine is voluntarily withheld long enough, urgency and dribbling from overflow incontinence may result.

The association between DES and constipation is well established, although whether these are two consequences of a single mechanism, or whether constipation physically contributes to DES is not clear. In either case, it is well established that relief of constipation alone may lead to major improvements in voiding symptoms, and that lack of attention to constipation makes it unlikely that the voiding problems will be relieved [4].

Physical examination in suspected DES is most important in terms of pertinent negatives. Careful examination of the back should be done to insure that there is no cutaneous evidence of an underlying spinal abnormality. Similarly, strength, tone, sensation, and reflexes in the legs should be assessed. Some perineal irritation and erythema is commonly seen as a consequence of urinary dribbling. Although routine rectal examination is rarely needed, hard stool may be felt if this exam is performed. Sometimes, hard stool can be palpated on abdominal examination.

There is confusion and some disagreement regarding the diagnosis of DES. Some believe that formal urodynamic testing is a sine qua non for establishing the diagnosis of DES [3]. Other clinicians believe that a careful history and physical examination is usually sufficient. There is no strong consensus on this point. Most agree, however, that previously recommended procedures such as voiding cystourethrograms and cystoscopy are rarely needed.

As for nearly any urinary tract symptom, a urinalysis on a first morning specimen is a helpful starting point. Since some children with DES are infected regularly, evidence of UTI (nitrite, leukocyte esterase, and bacteria) is not unexpected; its presence neither confirms nor contradicts the diagnosis of DES. On the other hand, lack of urine concentration or the presence of fixed proteinuria suggests that an underlying nephropathy needs to be pursued.

For most children with suspected DES, the only needed imaging is a kidney and bladder ultrasound, including a postvoid view of the bladder. Evidence of kidney scarring or significant hydronephrosis merits evaluation by a nephrologist or urologist, although both of these can occur as complications of DES. Although the utility of an abdominal plain film for establishing constipation is not clear, it is sometimes useful in demonstrating to the family something which they may not appreciate.

Treatment

Most behavioral voiding disorders can be managed in the primary care setting. A major first step in addressing any of these problems is a clear explanation to the family and child, including unequivocal reassurance that the disorder is benign. Whether explicitly stated or not, many parents will harbor concerns that their child's urinary symptoms are evidence of a serious underlying kidney problem, and potentially are harbinger of kidney failure, dialysis, or the need for transplantation. As discussed above, the diagnostic workup of each of these problems is quite straightforward. It is rarely necessary to obtain additional testing or imaging beyond that recommended. The primary care practitioner should avoid the temptation to engage in additional testing to "reassure" the family. Indeed, such

continuing testing may simply provoke additional anxiety that there is some uncertainty regarding the child's problem.

Similarly, it must be remembered that each of these disorders ultimately is self-limited. With no treatment whatsoever, all three of the above conditions can be expected to disappear gradually as the child matures. Thus, an important goal of therapy is to insure that the child is not emotionally (or even physically) scarred by attempts at managing a self-limited problem.

Functional polyuria may be the easiest of the behavior voiding problems to manage. Frequently, it seems to develop suddenly and disappear within a few days almost as abruptly, although some studies cite 6 months as the mean duration of symptoms. Therefore, early in the course of the condition, encouraging parents and teachers to ignore the frequent urination may be all that is needed [5].

Usually, children with functional polyuria do not display urinary frequency throughout the day and in all settings. Rather, there may be specific situations in which the behavior is seen most dramatically. Such an observation is a good starting point for a discussion with the child and family regarding potential sources of anxiety associated with these settings.

Rarely, functional polyuria becomes disabling, by preventing the child from participating in school, play, or other activities. It also may become so prolonged that medical attention is sought. In such situations, more formal behavioral management may be called for. Hypnosis, as described in Chap. 21, may be a useful intervention in such cases although there are currently no studies addressing this.

Primary nocturnal enuresis is likely the functional urinary tract disorder with the widest range of impacts on the child and family. Some children and families may look upon bedwetting as more of a nuisance than a serious problem, and not even bring it to the attention of a medical professional. In others, however, it can be the genesis of severe family discord and even long-term emotional effect on the child.

Most typical forms of "behavioral" management are unlikely to be effective for nocturnal enuresis. The conventional understanding of the disorder is that neurologic immaturity is etiologically important. Thus, such classic behavioral interventions as rewards ("star charts") or engaging the child in dealing with consequences (washing bed linens) would not be predicted to be useful in a condition beyond voluntary control.

Although outside the purview of this chapter, the vasopressin analogue desmopressin is widely employed in the management of nocturnal enuresis. Beyond its effect on reducing urinary volume, recent data suggest a central effect of this agent as well [6]. Although generally safe in its oral form, nasal desmopressin is no longer recommended for this indication after disturbing reports of symptomatic hyponatremia in children receiving it. Desmopressin is expensive, and there is no evidence that its effect is sustained after discontinuation [7].

The simplest behavioral intervention for nocturnal enuresis is simply awakening the child a few hours into sleep and taking him to the bathroom to void. Although seemingly common sense, the technique has recently been studied carefully in a controlled fashion and found to be helpful [8]. If a family has not already tried this (generally, most have prior to consulting a physician), it is a simple initial intervention. The same comments pertain to restriction of fluid intake after dinner.

The most widely employed "behavioral" intervention for nocturnal enuresis is the use of alarm systems. Far advanced from the cumbersome units of a decade or two ago, modern enuresis alarms consist of a small sensor placed inside the child's underpants, which is connected to an alarm about the size of a small pager. The system awakens the child (or, sometimes more often, the parents) at the first detection of moisture by the sensor. The child then completes voiding in the toilet. The decision to employ an alarm system requires full understanding of the parents that initially they will be crucial to its success. At least initially, they will be awakened and will need to respond [2].

When used properly, the success rate of enuresis alarms is about 30 % in some published studies and is higher in our experience. There is also

evidence that the combination of desmopressin and an alarm system may be useful when neither alone works.

The behavioral management of DES, while complex and time-consuming for all involved, can be extraordinarily satisfying, even life-changing, for the child and family. It does, however, require a significant investment in time and resources by both the medical provider and the family.

The management begins with education. The concept that DES is a learned behavior, which developed over a long period of time and which can be "unlearned," is crucial for success. It must similarly be stressed that although we refer to the condition as "learned" and "behavioral," the child's symptoms are certainly not volitional, nor should they be subject to punishment.

If the child has had the imaging recommended above, it may be useful to show this to the family during the educational process. The ultrasound image of a large bladder, which does not empty completely, is fairly easy to demonstrate and can be enhanced by contrast to a normal study. A plain film demonstrating significant constipation can be helpful, especially when the extent of this problem is not appreciated by the child or family.

The next step is establishing a regular routine for voiding. Children with DES need to be encouraged to attempt voiding about every 2–3 h during the day, at home and at school. The latter may be difficult if the child has difficulty using a public restroom; this may involve enlisting the school's help in identifying a private bathroom, such as that in a school nurse office [3].

The timed voidings must be unhurried and as minimally stressful as possible. Some girls with DES whose feet do not touch the floor while seated on the toilet may find it easier to void (and to defecate) if a small bench or stool is situated such that their feet rest upon it. Additionally, some girls (especially those with an element of obesity) may find voiding easier by sitting backward on the toilet, facilitating wide spreading of their legs. A voiding calendar, with rewards established for adherent days, is useful.

As efforts at increasing voiding frequency begin, a corresponding increase in fluid intake is advisable. Children who are accustomed to voiding twice per day may find it difficult to void with greater frequency in the absence of an increased daily urine volume. One way to achieve this is to incorporate drinking water after each void.

There are some data that the use of biofeedback techniques may be another modality to improve both bowel and bladder function in children with DES (see Chap. 20) [9].

Addressing constipation is critical to the success of treating children with DES. Establishing a time for a daily bowel movement, typically following a meal, is an important first step, and this can also be incorporated into the voiding calendar. Increasing dietary fiber is another important component of this, although the reality of the "typical" American diet makes this difficult. Fiber supplements are probably a more realistic way of accomplishing this. Most children whose DES is associated with significant constipation will require initial therapy with osmotic (e.g., polyethylene glycol) and stimulant (e.g., senna) cathartics [10].

For most children with DES, the above steps are very likely to bring about significant improvements. Children who do not respond to behavioral intervention alone may require referral to a pediatric nephrologist or urologist. Occasionally, anticholinergics, prophylactic antibiotics, or other medications may need to be added to the regimen. Even if this is the case, however, the above behavioral program remains the mainstay of treatment.

Case Studies

Case 1

Jamie is a 6-year-old who was seen by his pediatrician because of frequent urination. During the day, he would void in small amounts as often as every 15 to 20 min, although he could go on longer car rides without needing to void. Although he would occasionally wet the bed at night, he was dry 13 out of 14 nights. He did not drink an unusual amount of liquid.

Jamie's frequent urination developed a few days after starting first grade and caused some disruption at school. He had no dysuria,

urgency, or symptoms of urinary tract infection. There was neither significant past medical history nor any family history of renal disease.

The physical examination was unremarkable. A specimen of urine obtained upon awakening in the morning was negative for glucose, protein, and formed elements; the specific gravity was 1.021.

The history, exam, and urinalysis findings with Jamie are completely consistent with functional polyuria, and no additional testing is required.

Questions

1. The absence of a concentrating defect on this urinalysis is confirmed by which:
   (a) Absence of glucose
   (b) Absence of protein
   (c) Absence of formed elements
   (d) Specific gravity of 1.021
   (e) None of the above
2. Which of the following would argue against Jamie's polyuria being functional:
   (a) Maternal uncle with hypertension
   (b) Coincident new onset of nocturnal enuresis
   (c) Treatment for UTI at 9 months of age
   (d) Use of inhaled corticosteroid for asthma
   (e) Normal bowel movements
3. If Jamie's first morning urinalysis had demonstrated a specific gravity of 1.005, the appropriate next step would be:
   (a) A voiding cystourethrogram
   (b) A renal ultrasound
   (c) A urine culture
   (d) MRI of the brain
   (e) Questioning the circumstances of obtaining the specimen and repeating the test being sure that Jamie drank nothing between bedtime and the time of obtaining the urine sample

Answers

1. (d): 2. (b): 3. (e). The onset of frequent daytime urination with no change in nighttime frequency is characteristic of functional polyuria. A urine specific gravity of greater than 1.015 supports the absence of a concentrating defect. If the first morning urine is not concentrated, this finding should be verified on subsequent urinalyses, since children sometimes gain access to fluids during the night or prior to obtaining the morning sample.

Case 2

Rick is a 12-year-old boy referred by his pediatrician because of bedwetting. He was easily toilet trained for bowel and bladder during the day by 2 years of age. He had been hospitalized twice in the first year of life because of dehydration, but otherwise had been very healthy. Despite nighttime fluid restriction, he wets the bed completely about 6 nights out of 7. He once received a "pill" to control bedwetting, the name of which his mother does not recall, but this led to no change. The family history is negative for renal disease. Rick is in seventh grade, and doing well.

Rick's physical examination shows a height at the 5th percentile and weight at the 50th percentile. His blood pressure is normal. There are no abnormal findings.

The urinalysis was negative for glucose, blood, and formed elements; it was 1+ positive for protein, and the specific gravity of a first morning specimen was 1.008. The urinalysis was repeated on two other mornings, and these findings were confirmed. An ultrasound showed two normal-sized kidneys with increased echogenicity. There was no evidence of obstruction; the bladder was large, but emptied completely with spontaneous voiding.

At first glance, an otherwise well 12-year-old boy who has continually wet the bed but who has no daytime symptoms would be suggestive of primary nocturnal enuresis. The red flag here, however, is persistently dilute urine on many morning specimens. This suggests that a concentrating defect is responsible for the nocturia; the fact that he apparently has a generous bladder

capacity may have kept him from realizing that he was voiding a lot during the day. Episodes of dehydration early in life are seen in children with long-standing concentrating defects. The proteinuria is further evidence of a primary nephropathy. The normal-sized kidneys with increased echogenicity suggest some parenchymal involvement. A likely explanation of this scenario is nephronophthisis, an autosomal recessive disorder of the kidneys, which can progress to kidney failure in late adolescence or young adulthood [11]. Interestingly, some children with this disorder may have short stature, as does Rick. The "pill" he probably received previously was desmopressin, a vasopressin analogue which works in bedwetting by enhancing urine concentration. The drug does not work, however, if the kidney's have a primary problem with their concentrating ability.

Rick needs to be referred to a pediatric nephrologist for further study and treatment. His bedwetting is unlikely to respond to any behavioral interventions.

Questions

1. A renal ultrasound is not typically obtained in a child with suspected primary nocturnal enuresis. The test was done for Rick because:
   (a) He was over 10 years old
   (b) He had short stature
   (c) There was suspicion of urinary tract infection
   (d) He had an abnormal urinalysis
   (e) There was no family history consistent with primary nocturnal enuresis
2. Rick did not have daytime voiding symptoms because:
   (a) He reduced his fluid intake
   (b) He accommodated to an increased urine output by holding his urine
   (c) He had coexistent constipation
   (d) Proteinuria resulted in a decreased volume of urine when upright
   (e) He was in denial
3. Interventions which could reduce Rick's bedwetting include:
   (a) Biofeedback training
   (b) Chronic antibiotic use
   (c) Reduction in fluid intake
   (d) Hypnosis
   (e) None of the above

Answers

1. (d): 2. (b): 3. (e). This case nicely illustrates that although isolated nocturnal enuresis is rarely a sign of underlying nephropathy, it may be and the urinalysis is generally the clue. The presence of proteinuria and a concentrating defect were clues to a renal problem. Because Rick probably had been producing a large volume of urine all his life, he was able to accommodate to this during the day, and only had wetting when asleep. Because of his increased urine output, there may be little which can be done for his bedwetting, although nephrologists might provide a trial of a medication to relax the bladder further.

Case 3

Susan is a 6-year-old girl referred by her family physician because of daytime dribbling of urine. She had been toilet trained for bowel and bladder at about 3 years of age, although she always had difficulty passing her stools. About a year ago, her mother noted that she was holding her urine during the day, sometimes voiding only twice. She was dry at nighttime.

For several days before this visit, she was running to the bathroom to void, but often getting there too late and consequently dribbling some urine onto her underpants. She had also begun having bowel movements only every 4–5 days, and complained of quite a bit of pain with each. When not voiding, she was noted to periodically be crossing her legs and grimacing.

She was otherwise well and had never been hospitalized. There was no family history of renal disease. Her physical examination showed normal growth and blood pressure. There were no abnormal findings.

A urinalysis was negative for blood, protein, and formed elements. The specific gravity was 1.020. A renal ultrasound showed two normally positioned kidneys with no obstruction.

The bladder was smooth walled and capacious and did not empty completely after voiding. A plain film of the abdomen showed a considerable amount of stool throughout the colon. The vertebral bodies were intact.

Susan shows the characteristic features of DES. This learned behavior likely began around the time of toilet training, but did not become noticeable until recently.

Questions

1. The observed behavior by which Susan was standing with crossed legs and grimacing is likely explained by:
   (a) The development of a UTI
   (b) The development of acute vaginitis
   (c) An attempt to prevent bladder emptying
   (d) Pain from renal colic
   (e) Attention seeking
2. A finding on history or physical examination which would suggest a diagnosis other than DES in Susan would be:
   (a) A dimple over the sacral area with decreased lower extremity reflexes
   (b) Hypertension
   (c) Leukocytes and bacteria on urinalysis
   (d) Erythema and excoriation around the vulvar area
   (e) A family history of type 2 diabetes
3. Additional testing which *could* help to establish the diagnosis of DES in Susan would be:
   (a) A DTPA renal scan
   (b) Urodynamics
   (c) Urine electrolytes
   (d) Barium enema
   (e) Cystoscopy

Answers

1. (c): 2. (a): 3. (b). Susan is displaying the characteristic behavior of a child attempting to hold her urine, and with the rest of the history has classic DES. With the exception of the possibility of a neurologic lesion, the other observations in question 2 are either consistent with DES or unrelated. Some experts recommend urodynamics as part of the workup, but the other options in question 3 have no place in diagnosis.

Conclusions

The storage and emptying functions of the lower urinary tract are usually distinct from the hormonal and excretory functions of the kidney. Disorders of voiding typically occur in the absence of kidney functional disturbances. The diagnosis of most voiding dysfunctions is made by a careful history and physical examination. A urinalysis on a first morning specimen is often the only laboratory test required. Education, reassurance, and some simple behavioral interventions will address the bulk of voiding disorders in children. Consistency by parents and providers, along with realistic expectations, are critical to successful treatment.

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Functional Symptoms in Pediatric Dermatology: The Canary in the Coal Mine

7

Anna J. Nichols and Ted A. Grossbart

Abstract

The skin is a window into the state of health revealing genetic predispositions, hormonal balance, nutritional status, and internal wellness. It exposes stress burden, emotional well-being, and psychological health. In this way the skin can operate as the metaphorical canary in the coal mine announcing the body's hidden alarm signals. Psychocutaneous disorders draw attention to the intimate connections between the mind and the skin. During development, the brain and skin are born from the same embryonic tissue and throughout life they continue to be connected through elegant interactions involving neuropeptides, hormones, and inflammatory mediators, the so-called molecules of emotion. Disfiguring skin disease such as vitiligo, acne vulgaris, and ichthyosis have large emotional and psychological interactions. Perhaps surprisingly, the extent of the psychological fallout that results from these conditions is not necessarily related to the objective nature of the disease. Instead it is highly correlated with its subjective severity, which is dependent on self-perception and is highly personal. Recurrent skin disease often results in anxiety, depression, social isolation, body dysmorphic disorder, and dramatic reductions in quality of life rivaling those associated with debilitating systemic illnesses. Crosstalk between the mind and skin also exists wherein physical or psychological stressors can both initiate and worsen skin disease that has clear organic underpinnings such as acne vulgaris, psoriasis, atopic dermatitis, alopecia areata, and cutaneous warts. Primary psychiatric disorders may result in skin and hair conditions such as

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trichotillomania, psychogenic excoriation, dermatitis artefacta, and delusions of parasitosis. Although mind–skin interactions can cause obstinate symptoms and skin disease, these same connections can also be harnessed for therapeutic benefit.

Keywords

Acne vulgaris • Atopic dermatitis • Pruritus • Psychocutaneous disorders • Psychodermatology • Quality of life • Skin • Stress • Trichotillomania • Warts

Background: Literature Review

The skin, the largest organ in the body, is a tactile sensor in an ever-changing environment. To sense and respond to the world, the skin is equipped with a dense network of specialized nerve fibers that enable communication with the central nervous system, the immune system, and the endocrine system. The skin provides touch, itch, pain, pressure, position, and temperature sensory input to the central nervous system. In response, the central nervous system controls blood flow to the skin, function of the smooth muscle surrounding individual hair follicles, and sweat response. Also, it influences immune responses and regulates hormones that directly affect the skin.

Several studies have shown that somatization is pervasive in patients with dermatologic disease, perhaps partly because they also tend to have elevated levels of anxiety related to their health [1–3]. Psychocutaneous disorders which are situated at the intersection of dermatology, psychiatry, and psychology illuminate the integration of the mind and body.

Chronic stress results in the dysregulation of immune system function. The stress response results in the release of neuropeptides and hormones that serve as the brain's alarm signals to the body. The physiological purpose of an acute stress response may be to prime the immune system in preparation for an imminent challenge. In a state of chronic stress, the over-activated immune system can target innocuous or self-antigens and result in bodily mayhem [4, 5]. Chronic stress may exacerbate inflammatory and autoimmune skin disease by reducing the number of leukocytes, suppressing leukocyte trafficking and function, or changing cytokine balance [6–8].

Significant differences exist among individuals in the way in which stress is perceived, processed, and ultimately managed. "Functional" symptoms can be thought of as physical manifestations of these alarm signals. If emotional and psychological stressors are not processed and coped with effectively, the energy that results can be directed inward. How this energy manifests into physical symptoms is individual and can affect virtually any organ. When it disrupts the homeostasis of the skin, the symptoms that it produces are dependent on overall health, genetic predispositions, environmental factors, latent viruses, and psychosocial history [9]. A similar stressful event can trigger intense itching in one person and a painful rash, hives, warts, herpes, or acne in others.

The condition of the skin has a profound impact on overall body image. Skin disease is rarely life-threatening, but often results in significant suffering that is out of proportion to the medical severity of the condition. Skin disease is inherently more difficult to hide than other afflictions. Patients affected by skin disease frequently suffer from anxiety, depression, low self-esteem, and social withdrawal. While suicide among patients with skin disease is relatively rare, self-destructive behaviors abound. Examples include quitting a sports team, abandoning a hobby, isolating oneself from friends and family, or non-adherence with medical therapy.

The connection between stress and its ability to cause or exacerbate skin disease is not a novel

concept [10–13]. Dr. Robert Griesemer, a pioneering dermatologist at Harvard Medical School, developed a method for looking beyond the skin [10]. He would ask his patients about conflict in their lives. Then he would compare their emotional well-being with their skin symptoms. From these comparisons he developed the Griesemer Index, a catalog of skin symptoms, which includes the percentage of cases that are triggered by emotional stress and how quickly stress results in symptoms (Table 7.1).

Psychocutaneous disorders can be divided into three principal categories, although there is frequently overlap between the groups [14].

1. Psychosomatic cutaneous disorders are caused by organic disease but can be triggered or exacerbated by psychological stressors; these include atopic dermatitis, alopecia areata, psoriasis, acne vulgaris, and warts.
2. Primary psychiatric disorders are responsible for self-destructive skin conditions such as trichotillomania, psychogenic excoriation, acne excoriée, body dysmorphic disorder, dermatitis artefacta, and delusions of parasitosis.
3. Secondary psychiatric disorders result from disfigurement associated with skin disease such as vitiligo, acne conglobata, and ichthyosis.

The extent of the psychological fallout resulting from skin disease is highly individual and most often based on the subjective assessment of severity of the disease. In the sections that follow, mind–skin connections will be explored using atopic dermatitis, trichotillomania, and acne vulgaris as classic and relatively common examples.

Table 7.1 Dermatoses with highest incidence of emotional triggering

Diagnosis	Percentage of diagnoses triggered by emotions	Biologic incubation interval between stress and clinical change
Hyperhidrosis	100	Seconds
Lichen simplex chronicus	98	Days
Neurotic excoriations	98	Seconds
Alopecia areata	96	2 weeks
Warts, multiple and spreading	95	Days
Rosacea	94	2 days
Pruritus	86	Seconds
Lichen planus	82	Days
Dyshidrotic hand dermatitis	76	2 days for vesicles
Atopic dermatitis	70	Seconds for pruritus
Factitial dermatosis	69	Seconds
Urticaria	68	Minutes
Psoriasis	62	Days
Traumatic dermatitis	56	Seconds
Dermatitis, not otherwise specified	56	Days
Acne vulgaris	55	2 days for papules
Telogen effluvium	55	2–3 weeks
Nummular dermatitis	52	Days
Seborrheic dermatitis	41	Days
Herpes simplex and zoster	36	Days
Vitiligo	33	2–3 weeks
Pyoderma and bacterial infections	29	Days
Nail dystrophy	29	2–3 weeks
Epidermal cysts	27	2–3 weeks
Warts, single and multiple	17	Days
Contact dermatitis	15	2 days
Fungal infections	9	Days
Basal cell carcinoma	0	Not applicable
Seborrheic keratoses	0	Not applicable
Nevi	0	Not applicable

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Atopic Dermatitis

Atopic dermatitis is a recurrent inflammatory skin condition characterized by a chronic course, severe pruritus, and frequent exacerbations. The pathophysiology of atopic dermatitis includes epidermal barrier dysfunction and dysregulations of the immune system [15]. In genetically predisposed individuals biological, psychological, and social factors interact and modulate the onset and clinical course of the disease. Triggers, both exogenous and endogenous, are numerous and

their contributions differ with individual susceptibilities. The result of these factors is that the morphological features and the course of the disease are diverse and variable.

Atopic dermatitis usually develops in childhood, often by 2 years of age. Estimates suggest that around 20 % of all children are affected to varying degrees [16, 17]. Because of skin's function in sensory perception and communication, infants with atopic dermatitis can have substantial and enduring ramifications in terms of emotional development [18–20]. Most cases of atopic dermatitis are mild; however, roughly 25 % result in moderate to severe reductions in quality of life [21].

Parents consistently rate the quality of life reductions in moderate to severe atopic dermatitis as similar to that of asthma but significantly worse than that due to diabetes [22]. Others have shown that atopic dermatitis has a more substantial impact on quality of life than enuresis, diabetes, asthma, and cystic fibrosis [23].

Children rate the severe, intractable itching and the resultant sleep disruption as the most important negative factors in their disease [23, 24]. Loss of sleep due to prolonged sleep latency and frequent awakenings causes both mental and physical exhaustion. Consequences of this include fatigue, mood and behavior problems, strained relationships, impaired concentration, and poor school performance [25–27]. Loss of sleep, especially during exacerbations, affects not only the patient but also siblings and parents [28]. This is significant because loss of sleep has been correlated with increased parental anxiety and depression [29].

It is well established that emotional and psychological factors play important roles in the etiology and clinical course of atopic dermatitis [14, 30–32]. Dysfunctional parent–child relationships have been noted in cases of intractable atopic dermatitis but it is not clear whether this is a cause or an effect of the skin disease [33]. The largest study investigating the effects of stress on atopic dermatitis was performed in the Japanese city of Kobe after a devastating earthquake in 1995. In the region most directly affected by the disaster, 38 % of patients had an exacerbation of their atopic dermatitis in contrast to only 7 % in areas that were spared by the earthquake [34]. Interestingly, patients with atopic dermatitis express significantly more extracutaneous psychosomatic symptoms when compared to the general population [35].

Although stressors in isolation do not likely cause atopic dermatitis, they can play an essential role in precipitating and exacerbating the disease. Exacerbations of atopic dermatitis may be accompanied by emotional and psychosocial reactions, which in turn can affect the disease process itself. One theory is hypothalamic-pituitary axis dysfunction. When exposed to stressors, children with atopic dermatitis produce significantly less cortisol, which can increase inflammation [36]. Research is only beginning to shed light on the enigmatic mechanisms underlying these effects, but it appears that they alter epidermal permeability.

Psychological effects such as poor self-esteem, social phobia, school avoidance, isolation, anxiety, and depression are twice as prevalent in children with atopic dermatitis compared to controls and these effects are directly correlated with the objective severity of disease [14, 37]. Childhood and adolescence are psychologically vulnerable periods of time when skin disease can create self-consciousness, embarrassment, and strong feelings of inadequacy.

An intriguing study showed that parental psychological health, a supportive home environment, and low influence of the child's skin condition on family functioning predicted lower levels of anxiety, depression, and social withdrawal in the affected children [38]. Thus, the family unit may modulate the psychological fallout from atopic dermatitis.

A characteristic feature of atopic dermatitis is the itch–scratch–itch cycle. This cycle is often and aptly referred to as "vicious." It plays an essential role in the maintenance of the clinical manifestations of the disease. Scratching leads to momentary reductions in itching so that scratching behavior is reinforced by its relieving effects. Scratching also increases itching because it produces physiologic changes in the skin including inflammation, which lower the itch threshold,

result in greater levels of itch, and produce more aggressive scratching.

Correlations between itch intensity and emotional stress are well established. Itch can be directly elicited in individuals with atopic dermatitis by confronting them with personal and stressful events from their past [39]. In addition, life-altering events such as divorce increase the incidence of atopic dermatitis [40]. The underlying mechanisms for this may involve lowering the itch threshold or aggravating itch sensitivity. Histamine, vasoactive neuropeptides, and inflammatory mediators are released as a result of stressors. Additionally, the hemodynamic changes resulting from stress (modifications of cutaneous blood flow, alterations in skin temperature, and changes in sweat response) likely contribute to this cycle.

The chronic nature of atopic dermatitis coupled with frequent and seemingly unpredictable exacerbations suggests to patients that they have no influence on the course of their disease. This can engender feelings of resignation, helplessness, and hopelessness and affect adherence to the prescribed treatment regimen.

Making the diagnosis of atopic dermatitis is generally straightforward. Alternate diagnoses that should be considered include contact dermatitis, acrodermatitis enteropathica, herpes simplex virus infection, *Staphylococcus aureus* infection, scabies, hyperimmunoglobulinemia E (Job) syndrome, phenylketonuria, and Wiskott–Aldrich syndrome. As with all psychocutaneous disorders, determining the contribution of emotional and psychological issues to the onset and course of disease can be challenging.

Trichotillomania

Trichotillomania is an impulse control disorder. It is on the spectrum of obsessive–compulsive disorders and some conceptualize it as a form of addiction [41]. It is characterized by a compulsive and habitual urge to remove one's hair and is preceded by a feeling of heightened tension and anxiety. Relief, relaxation and even pleasure, albeit momentary, follow the act of hair pulling [42]. Pulling becomes reinforced by this cycle and over time the behavior can become habit. It most commonly presents during childhood and adolescence and is a relatively common cause of alopecia in childhood [43]. In young children trichotillomania may simply represent a habit. In adolescence, however, it usually signifies a more serious psychological disorder. The prevalence has been estimated to be roughly 4 % of the general population with perhaps 10 % having been affected at some point during their lives [44]. The actual prevalence of trichotillomania among children and adolescents has been estimated to be around 1 % [45, 46].

The scalp is most often affected, often biased toward the side of handedness, but eyebrows, eyelashes, and pubic hair may be targeted as well. A continuum of hair pulling behavior exists that ranges from a relatively benign form that produces no distress to a serious disorder that is disfiguring and produces significant emotional and psychological suffering. Trichophagy is often present and can lead to the formation of a trichobezoar ("hair ball"). This so-called Rapunzel syndrome may result and can lead to anemia, ulceration, small or large bowel obstruction, intussusception, and perforation [47].

Individual insight into trichotillomania is variable. Children often engage in hair pulling during emotional and psychological duress. One theory proposes that pulling provides a stimulus of "counter irritation" that distracts the child from feeling dysphoria and anxiety [45]. Although there exists significant differences in pulling behavior, trichotillomania is not usually a conscious, focused act [43, 48]. Instead individuals tend to pull in "trance-like" or semiconscious states. It has been said that the hands of individuals with trichotillomania appear to "have a mind of their own."

Trichotillomania can be disfiguring and like other psychocutaneous disorders it is associated with a heavy emotional and psychological burden [49]. The duration of hair pulling appears to partially dictate the clinical course. Patients who pull for less than 6 months tend to have a relatively benign and short-lived course, whereas those who pull for 6 months or longer tend to

have a chronic and often treatment-resistant course [50, 51]. When it becomes chronic, quality of life is drastically reduced. Anxiety and depression are pervasive [52]. Body dysmorphic disorder, feelings of inadequacy, and perfectionistic personality types are highly prevalent as well. Attention-deficit and obsessive–compulsive disorders may be present [53]. These may contribute to the expression of the illness and often also intensify as a result of trichotillomania in this unfortunate cycle.

The task of diagnosing the cause of hair loss can be difficult for clinicians. The diagnostic possibilities, however, are narrowed substantially in healthy children with previously normal hair. Inquiring about other illnesses, stressors, and family history is essential. The diagnosis of trichotillomania can be made clinically in the vast majority of cases, thereby eliminating the need for additional investigations such as bacterial or fungal cultures, blood tests, or scalp biopsies. The suspicion for trichotillomania should be raised if there is a history of depression, anxiety, or obsessive–compulsive behavior. Diagnostic considerations include tinea capitis, traction alopecia, alopecia areata, loose anagen syndrome, telogen effluvium, monilethrix, and secondary syphilis. A careful history will elucidate important characteristics about the current episode of hair loss and any previous episodes [54]. It is important to examine for signs of anemia and thyroid disease. Clinical examination should focus on scalp skin, nails, body, and facial hair. In cases of trichotillomania, the affected areas are never completely devoid of hair. Dermoscopy reveals short, broken hairs often in bizarre, irregularly shaped patches with angular or linear borders [55]. Excoriations, hemorrhages, or perifollicular erythema can be present.

Acne Vulgaris

Adolescence is a time in which peer acceptance is critical. It is also a time when people tend to be extraordinarily self-conscious about physical appearances. Acne vulgaris is an extremely common problem, affecting more than 85 % of adolescents to varying degrees [56]. Acne is one of the most frequently encountered conditions in dermatology despite the estimation that only 20 % of affected individuals actually seek treatment from physicians. The development of acne during infancy and early childhood is often a forerunner of severe acne in adolescence [57]. Acne is a multifactorial disease. The physiological sequence of events underlying the disease process includes altered keratinization within the pilosebaceous unit, microcomedo formation, increased sebum production, proliferation of resident bacterial species including *Propionibacterium acnes*, and perifollicular inflammation. Genetics, hormones, diet, and psychological factors play a role in the onset and clinical course of acne [58–64].

Emotional and psychological factors have been identified as causative factors in acne and may impact acne in several distinct ways [64–67]. For example, emotional stress can initiate and/or exacerbate acne, psychiatric illness can result from the emotional and psychological scars left by acne, and primary psychiatric illnesses such as body dysmorphic disorder and obsessive–compulsive disorder can develop based on complaints centered on acne. Patients often describe a temporal relationship between periods of increased stress or anxiety with exacerbations of acne [10, 68, 69].

Several studies of adolescents have shown a positive correlation between acne severity and stress levels [63, 64, 66, 67]. In one study intense and stressful interviews produced a quantitative increase in acne lesions within days of the experimental stressor [70]. The latency period for the development of lesions was approximately 2 days, which is in agreement with that reported by Griesemer (Table 7.1). Additional evidence that stressors exacerbate acne comes from a small study that combined biofeedback relaxation and cognitive imagery in individuals with acne [71]. Over the 6-week study period, there was a significant reduction in acne severity in the biofeedback group compared to controls. Moreover, those that continued to practice the skills maintained the benefits, whereas those who did not failed to maintain their gains.

The biologic mechanisms underlying these effects are still unclear. One possible explanation is that hormones such as corticotropin-releasing hormone, which is a critical modulator of cutaneous stress responses, may alter the production of inflammatory mediators such as substance P. Hormones could also influence the specific lipid composition of sebum, thereby tipping the balance towards a proinflammatory state [63, 72–74]. The 70-year-old brain–gut–skin theory is an intriguing hypothesis that has gained support from recent preliminary studies. This theory proposes that emotional and psychological stressors alter gut motility, microflora profile, and intestinal permeability. This sequence of events allows toxins to gain systemic access, ultimately resulting in systemic and local skin inflammation [75, 76]. In predisposed individuals, this inflammation could then manifest as acne. In individuals without a strong predisposition toward acne, this same cascade could produce different chronic inflammatory conditions.

The emotional and psychological effects of acne can be significant and enduring. It can lead to social isolation, difficulties with interpersonal relationships, mood disorders, poor self-esteem, anxiety, social phobia, and depression [77–80]. In severe cases, it can increase suicidal ideation and suicide attempts, even after controlling for the presence of depression and anxiety [81, 82]. Improvement of acne is associated with improvements in psychological factors and quality of life [57, 83]. In a study of 1,566 adolescents, the psychological impact of acne was greater than or equal to that of patients with psychiatric disorders, epilepsy, asthma, diabetes, back pain, rheumatic pain, cystic fibrosis, and cancer [84, 85]. In that study, only patients with cardiac disease had greater impairments.

It had been previously thought that only patients with severe and objectively disfiguring acne such as acne fulminans or acne conglobata experienced significant psychological distress [30, 86–88]. We now know that individuals with mild to moderate acne report psychological distress that is comparable to those with severe acne [89].

There is a direct correlation between the *subjective* severity of acne and the presence of anger, anxiety, shame, loneliness, depression, and low self-esteem [81, 90, 91]. Excepting suicidal ideation and attempts, the psychological fallout of acne does not appear to directly correlate with the *objective* severity of the disease. In light of this, it is not surprising that body dysmorphic disorder is present in approximately 20 % of acne sufferers [92]. This condition creates a drastically distorted, negative view of their appearance, causing them to go to extreme lengths to fix problems that only they see.

The diagnosis of acne can typically be made from across a room by a trained clinician. However, very similar acneiform eruptions may result from infections, drug reactions, genetic disorders, and hormonal or metabolic abnormalities. Alternate diagnoses to consider include perioral dermatitis, folliculitis, acne conglobata, acne fulminans, rosacea, sebaceous hyperplasia, syringoma, and adenoma sebaceum in tuberous sclerosis. Acne excoriée and body dysmorphic disorder are important diagnostic considerations, as are anxiety and depression. Assessing the impact of emotional and psychological factors on the onset and clinical course of acne is challenging. There is currently no lab test or biopsy result that can parse out whether the lesions that we observe result purely from stress, biology, or a tangled combination. While many cases of acne are straightforward to treat, in many others the standard therapeutic arsenal will be largely ineffective without addressing the biopsychosocial dimensions.

Treatment

Despite the high prevalence of psychocutaneous conditions, the majority of skin conditions are caused by organic processes. Additionally, serious systemic disease can initially present with cutaneous manifestations, so all patients must have a comprehensive evaluation. Medical or surgical therapy is sufficient in some cases but is grossly inadequate in others.

It would be prudent to consider psychological factors in all patients with skin disease, but it is particularly imperative in those with chronic dermatoses. Additionally, the emotional and psychological contributions to skin disease should be closely examined when traditional medical therapies have been ineffective, when skin symptoms worsen during times of duress, and when skin disease causes significant reductions in quality of life.

Despite the prevalence of psychocutaneous disorders, most dermatologists are not familiar with or comfortable treating them [93]. According to one study, dermatologists failed to detect clinically significant anxiety and depression in many of their patients. Perhaps more surprisingly, even when they did successfully identify anxiety and depression, their treatment plans did not address these psychological impairments [94]. Results from surveys of both dermatologists and psychiatrists supply an explanation for this; there is a systematic lack of knowledge about psychocutaneous disorders [93, 95]. Only 18 % of dermatologists and 21 % of psychiatrists described a clear understanding of psychodermatology with the majority having little to no knowledge about the management of such conditions. The ramifications of these findings are compounded by the fact that patients frequently resist referral to psychiatrists or psychologists.

For some skin conditions, psychological therapies cannot cure the underlying cause, but can still offer substantial relief. Take liver disease as an example. There is nothing psychological about it or the intense itching that it often causes. But hypnosis and other psychodermatology tools can help people with liver disease experience less itch. Their symptoms improve even though the disease process is unchanged. There may be other benefits. Research has also found hypnosis helpful for people who have suffered severe burns. They heal faster, feel less pain, and are less likely to develop infections and other complications compared with burn patients who receive only medical treatment [96–99].

The ideal approach to patients with psychocutaneous conditions employs a biopsychosocial model to truly understand the individual and their skin condition. The most effective therapeutic strategies for such conditions are holistic and multidisciplinary. The psychodermatology therapeutic toolbox provides many options that support healthier behaviors, modulate stress responses, and can be tailored for individual patients. Rigorous evidence supporting their use is still largely lacking, in part because the nature of many of these therapies makes it impossible to evaluate them in double-blind randomized placebo-controlled trials.

Hypnosis (Chap. 21) employing ego strengthening and direct suggestion can help patients first identify and then reduce behavioral or psychological barriers to healing [100, 101]. Hypnosis was found to significantly reduce pruritus, scratching behavior, sleep disruption, tension, as well as frequency of topical steroid use in atopic dermatitis [102]. Additional therapies that have utility include psychotherapeutics (Chap. 26), breathing and yoga (Chap. 25), and meditation (Chap. 24). A recent pilot trial of acupressure (Chap. 23) at one specific acupoint showed a significant reduction in pruritus and lichenification resulting from chronic scratching [103].

Psychosomatic hypnoanalysis has been used to successfully treat recalcitrant cutaneous warts and is an excellent option especially for young children [104–106].

Cognitive behavioral therapy (Chap. 19), biofeedback (Chap. 20), and imagery (Chap. 22) have been shown to reduce the objective number of lesions in acne vulgaris, gains that were lost when the study participants stopped practicing these techniques [71].

A strong physician–patient relationship is critical when treating patients with trichotillomania. Insight in this disease is highly variable. Children frequently deny hair pulling and direct confrontation is rarely useful. The impairment is semiconscious and over time the patient can often admit their role in manipulating the hair. If a specific psychosocial trigger can be identified, individual or family therapy can be useful. Psychotherapeutics (Chap. 26) including selective serotonin reuptake inhibitors, tricyclic antidepressants, stimulants, and antipsychotics may be useful in some cases, although a recent systematic review showed that habit-reversal therapy was the superior approach [107–109].

Case Studies

Case 1: Warts

SE was a 17-year-old healthy, female, high school senior who had been a ballet dancer since early childhood. She put her heart and soul into ballet, but wasn't sure if she wanted to continue it beyond high school because the odds of becoming a professional ballerina were so slim. She presented with multiple spreading plantar warts on the bottom of her right foot so painful that they prevented her from dancing. The warts were recalcitrant. She had tried countless topical destructive therapies. Each would eliminate the warts for a while, but then they would return. She had all but given up on the notion that she could ever be free of them.

Questions

1. Which of the following is the most effective topical therapy for warts?
   (a) Salicylic acid
   (b) Gentle cryotherapy
   (c) 5-FU
   (d) Imiquimod
   (e) Anthralin
2. At this point, which of the following is the most appropriate therapy for SE?
   (a) Systemic acyclovir to prevent human papillomavirus (HPV) shedding
   (b) Biopsy to verify the diagnosis of verruca vulgaris
   (c) Psychosomatic hypnoanalysis
   (d) Tylenol for pain reduction
   (e) Gardasil
3. What are possible explanations for the recurrence and spreading of warts in SE's case despite adequate medical therapy?
   (a) Poor patient compliance
   (b) Infection with a virulent HPV serotype
   (c) Lack of systemic immune response targeting HPV
   (d) Ambivalence about continuing with ballet
   (e) (c) and (d)

Answers

1. (a): Salicylic acid. Spontaneous regression of cutaneous warts occurs frequently, so benign neglect is a reasonable initial option. However, recalcitrant, recurrent, spreading, and painful warts warrant treatment. Treatment is difficult, destructive, and nonspecific with low cure rates and high recurrence rates. Greater than 40 unique topical therapies have been studied in randomized controlled trials. A recent meta-analysis reviewed the data from 77 trials of medical therapy for warts and concluded that the best evidence supports the use of salicylic acid. The cure rates in the reviewed trials were 23 % for placebo, 52 % for salicylic acid, and 54 % for aggressive cryotherapy. Gentle cryotherapy was no better than placebo. The combination of aggressive cryotherapy and salicylic acid was slightly better than either alone with a cure rate of 58 % [110, 111].
2. (c): Psychosomatic hypnoanalysis. This treatment modality has been shown to be an effective cure for warts. In addition this therapeutic option is painless and without side effects. Although the underlying mechanisms have not yet been revealed, an effect on the immune system is presumed.
3. (e): Both (c) and (d). Unlike infection with other viruses, HPV does not cause a generalized illness and does not incite a systemic immune response. HPV replicates in the living lower epidermis in a zone referred to as the stratum spinosum. In cutaneous warts, the virus tends to reside in the most superficial layer of the skin, the stratum corneum. The dormant form of the virus is shed from this layer. The stratum corneum is anuclear and devoid of a vascular supply, which enables the virus to evade surveillance by the immune system. The warts in this case, in effect, resolved SE's ambivalence about continuing with ballet. Under psychosomatic hypnoanalysis guided by a psychodermatologist, she conjured images of the virus that was causing her warts and she visualized her immune system attacking it. She envisioned her foot

clear of the warts. In therapy, she also recognized her ability to make decisions using her heart and her mind, so that she could leave her skin out of the decision-making process. SE came to understand that her skin was giving voice to something that she could not admit to herself and that she did not really want to pursue dancing professionally. After 3 weeks, her warts disappeared for good. SE's emotional stress may have weakened her immune system, tipping the balance in favor of the virus and making her susceptible to relapse. Our knowledge of the precise mind–skin pathway in cases like this is in its infancy. But clearly SE's warts were medical *and* psychological, and she needed medical and psychological treatments to cure them.

Case 2: Atopic Dermatitis

JO was an 8-year-old female with relentless atopic dermatitis, numerous allergies, and severe insomnia. Relapsing episodes of intense pruritus and extensive excoriations left her skin raw, bleeding, and vulnerable. She scratched, clawed, and dug into her skin until blood flowed. Nighttime was the most difficult. Without the day's distractions the urge to scratch was unbearable. Despite the best efforts of countless doctors, her condition was unresponsive to topical corticosteroids, calcineurin inhibitors, phototherapy, and systemic therapies. As years passed the scratching unabatedly devastated and scarred her skin and tortured her mind with feelings of shame, isolation, and hopelessness.

Questions

1. Which of the following are frequently comorbid with atopic dermatitis?
   (a) Asthma
   (b) Allergic rhinitis
   (c) Depression and anxiety
   (d) Low quality of life
   (e) All of the above
2. What might explain JO's persistent lack of response to standard therapies?
   (a) Inaccurate diagnosis
   (b) Unresolved emotional and psychosocial issues
   (c) Fatty acid deficiency
   (d) Malingering
   (e) All of the above
3. What additional therapies could be effective for JO?
   (a) Hypnosis
   (b) Psychotherapy
   (c) Biofeedback
   (d) Behavioral therapy
   (e) All of the above

Answers

1. (e): All of the above. The well-known association between atopic dermatitis, allergic rhinitis, and asthma is often referred to as the atopic march. Numerous studies have documented increased anxiety, depression, and reduced quality of life in patients with atopic dermatitis. Given the high prevalence of this condition and the presence of these comorbidities, the personal and societal costs are extraordinarily high.
2. (b): Unresolved emotional and psychosocial issues. Stress resulting from unresolved emotional and psychosocial issues can exacerbate cutaneous disease including atopic dermatitis, psychogenic excoriations, acne, psoriasis, urticaria, and alopecia areata (Table 7.1).
3. (e): All of the above. Hypnosis, psychotherapy, biofeedback, and behavioral therapy may be effective in targeting the causes for JO's unremitting itch in the face of escalating medical therapy.

Case 3: Acne

KT was a 15-year-old male who had been evaluated by many well-respected dermatologists for his severe, scarring nodulocystic acne, but the standard therapies made very little difference and only for a short period of time. His past medical history was remarkable only for stubborn infantile acne that resolved during infancy without medical intervention. During adolescence, he had

frequent exacerbations and with these his physical and psychological condition deteriorated. His self-esteem plummeted and he slowly traded social activities for solo pursuits. The state of his skin was never far from his mind. It affected his ability to concentrate at school and soon this distraction began to show in his grades.

Questions

1. Which of the following are true statements regarding acne vulgaris?
   (a) It is a monogenic disorder
   (b) It results from poor hygiene
   (c) It is purely a cosmetic concern
   (d) The psychological fallout is comparable to many chronic, debilitating diseases
   (e) Vaccination against *Propionibacterium acnes* dramatically reduces disease severity
2. What are some of the possible reasons for poor treatment outcome in KT's case?
   (a) Poor adherence to the prescribed treatment regimen
   (b) Untreated emotional and psychological factors
   (c) Self-defeating behaviors
   (d) Personal history of severe infantile acne
   (e) All of the above
3. What should the physician do next?
   (a) Screen for suicidal ideation and previous suicide attempts
   (b) Employ a biopsychosocial model to understand more about KT and his acne
   (c) Teach him mind–body techniques such as biofeedback and hypnosis
   (d) Consider referral to a psychiatrist and/or psychologist
   (e) All of the above

Answers

1. (d): The psychological fallout resulting from acne vulgaris is comparable to many chronic and debilitating diseases including asthma, diabetes, epilepsy, and cancer. The condition of the skin contributes significantly to body image and self-esteem. Individuals with acne can suffer tremendously and that suffering often endures long after the physical scars have healed. Acne is multifactorial, not monogenic. A myth from the past was that poor hygiene caused acne; we now know this to be untrue. The immune response to *Propionibacterium acnes* has been implicated in the pathogenesis of acne. Component vaccines and inactivated whole bacteria vaccines are being studied as possible new therapies for acne but they are not available [112, 113].
2. (e): All of the above are possible explanations for poor treatment outcome. Depressed mood, poor self-esteem, or anxiety that often result from acne can promote self-defeating behaviors such as poor adherence to the prescribed treatment regimen. Infantile acne is a frequent harbinger of stubborn acne in adolescence. Hidden emotional and psychological stressors can worsen acne and prevent remission despite appropriate medical therapy. Like a fallen tree that is blocking traffic, KT's distress is impeding the success of standard, usually reliable medical therapy, and must be addressed.
3. (e): All of the above. Some cases of acne are straightforward, but in others the medical treatments will be wholly ineffective without simultaneously targeting the underlying psychosocial factors. In this case, a comprehensive history addressing biopsychosocial issues is the immediate next step. Suicidal ideation and suicide attempts are much more prevalent in patients with severe acne. Clinicians in dermatology and primary care can learn mind–body techniques such as biofeedback and hypnosis that can be invaluable in the healing process. Referral to a psychiatrist and/or psychologist knowledgeable about psychodermatology should be employed in a tactful manner if the clinician feels uncomfortable delving into the emotional and psychological factors in cases such as this.

Conclusions

Stress is ubiquitous in our society and children are not insulated against its negative effects. The skin reacts directly to emotional stimuli and sometimes reveals secrets that the mind tries to conceal. It blushes when we are embarrassed and

perspires when we are nervous. The skin expresses fear, frustration, anger, and shame. It cries, rages, remembers, and punishes for real and imagined sins. Skin disease can function as a metaphorical canary in the coal mine declaring unresolved emotional and psychological issues. Stress, anger, relationship conflicts, and various psychological disorders can induce skin symptoms, and conversely chronic skin symptoms can lead to depression, anxiety, social withdrawal, and other psychosocial problems. Skin symptoms can be early warning signs of emotionally induced damage within the body. For children and adolescents, these conditions can be transformed into powerful opportunities to look inside and sort out emotional issues before they inflict more damage. Although the intimate connections between the mind and the skin can cause skin symptoms that frustrate patients and clinicians alike, these connections can also be harnessed to reduce them. Psychodermatology practitioners treat skin the way a psychotherapist treats behavior, by learning how it responds to emotional and environmental stressors and helping to moderate those responses.

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Psychogenic Influences Associated with Allergic Disorders

8

Juan L. Sotomayor Jr.

Abstract

Most discussions of functional respiratory disorders that come to the attention of allergists are centered on the vocal cords and the lower respiratory tract such as related to asthma. Little clinical attention has been focused on functional issues related to the upper airway and specifically those associated with rhinitis conditions. In addition, the evaluation of food allergy frequently is complicated by functional issues. It is therefore important for clinicians to become aware of clinical nuances related to psychological influences that can complicate proper diagnosis and management of these conditions. Familiarity with the differential diagnosis of allergic, non-allergic, and vasomotor rhinitis is important in order to make an accurate diagnosis of these respective clinical entities. Psychogenic sneezing is an example of a common psychologically triggered symptom that mimics an allergic reaction. Other diseases such as fibromyalgia, multiple chemical sensitivity, and chronic fatigue syndrome can present with rhinitis and also are associated with psychological influences. Understanding the etiology of food allergies may not only be life saving but also directs the proper management of the disease state and ultimate resolution of symptoms in some cases. In this context, quality of life and psychological burden of patients with food allergies should be considered, including the effects of psychological conditioning on propagating psychogenic food allergies.

Keywords

Allergic or non-allergic rhinitis • Psychogenic or factitious sneezing • Chronic fatigue syndrome • Fibromyalgia • Multiple chemical sensitivity • Food allergies • Hygiene hypothesis

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Introduction to Rhinitis and Allergies

The nose is rarely thought of as a site for conversion disorders (characterized by the loss of or alteration in physical function resembling a physical disorder in the absence of organ dysfunction) or psychosomatic illness [1]. The healthy nose is an often underappreciated organ that becomes a source of concern once dysfunctional [2]. Many clinicians may feel that rhinitis conditions are trivial, but for the patients they can substantially affect their quality of life [3]. Proper recognition and understanding of the nasal pathophysiologic mechanisms will lead to more accurate diagnoses and provision of optimal therapy for these patients.

In addition to their function of olfaction, the nasal passages have been described as an efficient "scrubbing tower" for the respiratory tract and act as an "air conditioner" and "defender" for the lower respiratory tract. The term rhinitis translates simply into "inflammation of the nasal mucous membranes," without implication of its trigger. An allergic condition is one of the leading causes of rhinitis that is unassociated with infection, with an estimated prevalence of 9–42 % and perhaps rising [4]. Other allergic disorders include food allergies, anaphylaxis, conjunctivitis, asthma, eczema, and urticaria.

Proteins (and more rarely lipids or carbohydrates) that are typically noninfectious foreign substances and generally innocuous to the immune system, can become allergens that are disruptive to well being. Allergens that are of small particle size and aerodynamically suited (i.e., molds, pollens, and dust mite) can penetrate the mucous epithelial membranes and the submucosal membranes and come in contact with a variety of cells including mast cells, T helper (Th2) cells, dendritic cells, basophils, eosinophils, and antigen-presenting cells (APCs) that orchestrate a classic induction of IgE antibodies.

Allergies occur when a patient becomes sensitized to an allergen after an initial exposure followed by abnormal IgE production. Upon reintroduction of an allergen, sensitized individuals already have allergen-specific IgE antibodies attached to mast cells by high-affinity IgE receptors [5]. The classic crosslinking of adjacent IgE molecules mainly on mast cells by bivalent or multivalent allergens triggers a complex immunological process called a Type I early-phase hypersensitivity reaction. Mast cells have preformed chemical allergic mediators stored in granules (principally histamine) that are released into the microenvironment after "degranulation." The pathophysiological changes that are induced by abnormal amounts of these chemicals include vasodilation, vascular permeability, increased mucus and secretions, and stimulation of sensory nerves. Most notably and importantly, the clinical reactions in the nose are exhibited by sneezing, congestion, rhinorrhea, postnasal drip, headache, and itching (Fig. 8.1). If the allergic reaction simply stopped there, it would not be of major clinical severity for most patients. However there is a well-recognized second stage called the late-phase reaction that typically occurs 2–6 h later and can persist for days.

The exact reason why chronic allergic inflammation persists is unknown, but remodeling and injury of local tissue, neurogenic stimulation, and introduction of bacterial organism all may play a role. Susceptibility to allergic sensitization and chronic inflammation is based on a myriad of factors. Genetic susceptibility in families, pollution, tobacco smoke, allergen concentration, and exposure routes alone or in combination are all suggested as potential abetting factors. Depression, stress, and altered mood disorders have also been suggested as predisposing factors [6].

Allergic Rhinitis

Allergic Rhinitis (AR) traditionally has been divided into seasonal or perennial. Seasonal allergies occur in sensitized individuals with a temporal relationship to specific allergens during peak seasons, such as is seen with grass or ragweed pollen. Perennial rhinitis occurs year round in reaction to nonseasonal allergens such as dust mite or dander and can vacillate in intensity throughout the year.

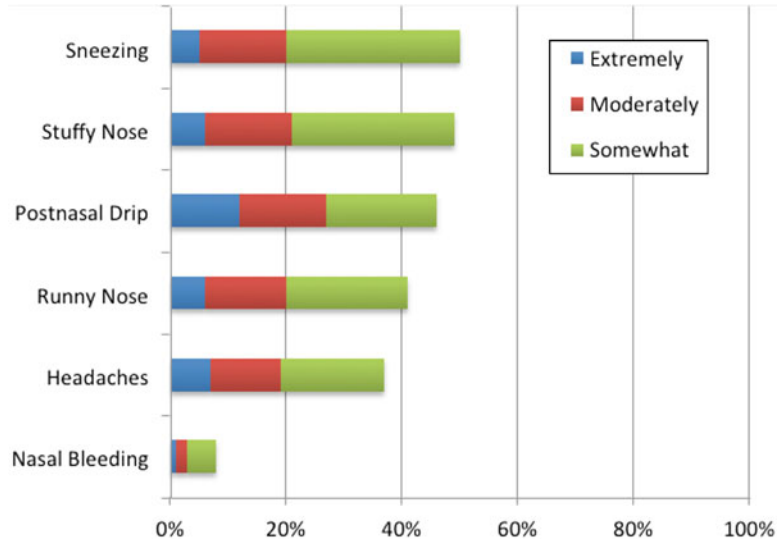

Fig. 8.1 Clinical reactions in the nose. Patients with nasal allergies were asked: How troubled have you been by these symptoms during the last week? Were you not at all troubled, hardly troubled, somewhat troubled, moderately troubled, or extremely troubled? (Adapted with permission from Meltzer [3]. With permission from Content Ed Net LLC)

Diagnosis of allergies is generally done through assessment of IgE activity by skin prick test, intradermal test, or various blood testing. Skin prick tests generally are done by placing a purified allergen on the skin and lightly abrading the skin with a sharp tip device. The size of any resultant wheal and flare reaction caused by an IgE response then is measured. Testing can be affected by numerous factors including prior antihistamine use, multivitamins, skin disorders, allergen extract potency, seasonality, and hypnosis [7]. The immediacy and safety of this technique are its major advantages. Blood allergy tests generally are felt to be less sensitive then the skin prick test.

All the aforementioned tests suffer from potential false positive and false negative results. False positive tests can occur in 8–30 % of asymptomatic patients, who do not suffer from a clinically recognizable allergic symptom even though the allergy test is positive. False negative testing which is less common occurs when a patient suffers from a clinical allergy but the testing does not detect abnormal IgE antibodies or IgE-mediated skin reaction [8].

These tests also suffer from another major drawback in that they reflect circulating IgE antibodies in the blood rather than in the nose. The best testing is direct inoculation of an allergen on the mucous membranes of the nose and measurement of localized IgE antibodies and subsequent inflammatory mediators, termed "entopy." This is not routinely done in clinical practice as the assays are difficult to run and not easily obtainable for clinicians. Up to 62 % of patients previously labeled as having nonallergic rhinitis or idiopathic rhinitis may in fact have only a localized nasal response to an allergen [4, 9].

AR and Psychological Influences

Nasal allergies can impact substantially a patient's quality of life. Nasal allergies can cause an emotional burden and are characterized by irritability, feeling miserable, depressed, anxious, or embarrassed. In addition, patients with AR can experience poor sleep, activity limitation, decreased productivity at work, and much fatigue (Fig. 8.2) [3]. These findings are not totally unexpected as rhinitis and an emotional link go back as far as the seventeenth and eighteenth centuries. "Rose fever syndrome" was described as an association of either the vision or odor of the rose and bizarre clinical reactions such as swooning or fainting [10]. There was a symbolic representation to these reactions, which may have represented femininity, weakness, and indecision. It must be noted

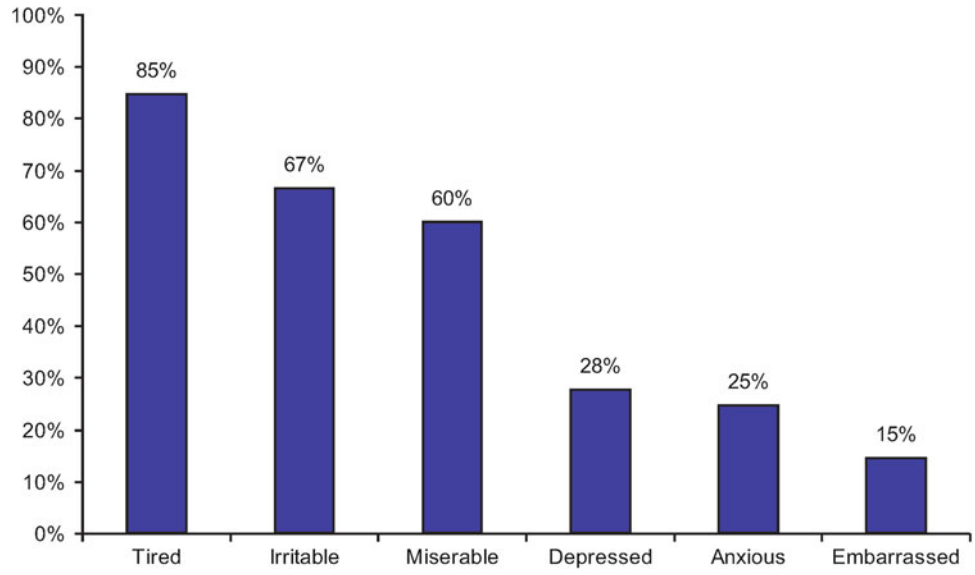

Fig. 8.2 Patient-related emotional burden during the worst month of symptoms ($N=400$). Patients with nasal allergies were asked: During the worst month of allergy symptoms, how often do you feel (depressed, irritable, tired, embarrassed, miserable, or anxious.)—frequently, sometimes, rarely, or never? (Adapted with permission from Meltzer [3]. With permission from Content Ed Net LLC)

that the typical reactions of rhinitis and conjunctivitis were recognized as probably organic in nature, yet these bizarre over-reactions were thought to be emotional in origin. As the nineteenth century approached, "Rose fever" was replaced by the term "hay fever" as other causes of allergens such as ragweed and dust became more recognized.

There is a long history and bidirectional relationship between psychiatric comorbidity such as depression and allergic rhinitis in the literature. The relationship is especially prominent in children [11]. Previous terms coined for psychiatric comorbidity include "allergic toxemia," "allergic irritability," or "allergic tension fatigue syndrome." These descriptive names typically share the following features: irritability, depression, fatigue, apathy, malaise, poor concentration, poor temper, and dysphoria.

Anxiety disorders are especially prominent in those with asthma and allergic rhinitis. Depression is seen more often in females with both AR and nonallergic rhinitis (NAR) Two large-scale studies suggested that patients with prior history of hay fever, who had positive allergy skin tests, and who received allergy shots were twice as likely to develop major depressive episodes during their midlife period. Identical twins have a much higher association of allergy and depression/anxiety than in the general population, which suggests a genetic link [12].

A number of studies have found that depression and poor mood (based on mood ratings scale) are clearly exacerbated during peak pollen periods such as seen with tree or ragweed pollen [13, 14]. Other studies have observed an increased incidence of suicide during peak pollen seasons in those with a history of allergy [15]. The authors suggest that allergy, mood disorders, and suicidal ideation may share underlying inflammatory and stress–response mechanisms as a possible etiologic link in their pathways.

Stress may induce a state of hyporesponsiveness of the hypothalamic pituitary axis, whereby cortisol secretion is attenuated. This leads to increased secretion of inflammatory cytokines that typically are counter-regulated by cortisol. Stress may also upregulate IgE responses, even in young children [6].

Fatigue and depression that are frequently seen with mood disorders are often associated with AR. The mechanisms involved are suggested

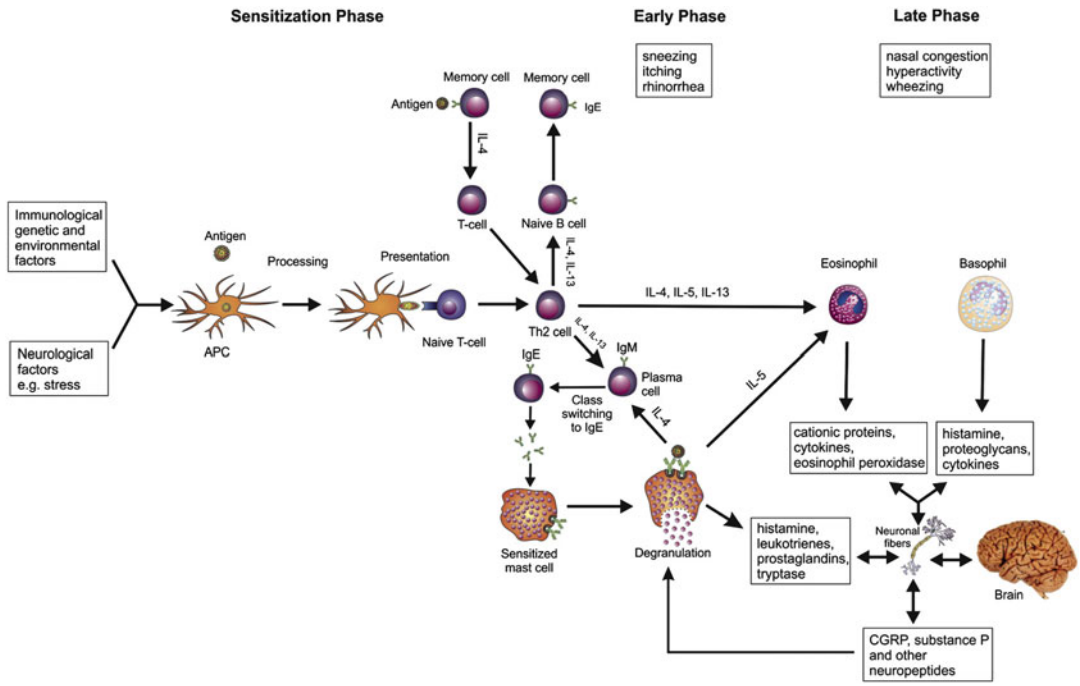

Fig. 8.3 Interaction of allergic rhinitis and stress. Schematic presentation of comprehensive mechanisms of AR. The figure depicts a highly simplified flow diagram of the complex process. Sensitization is a preparation phase where APCs encountering allergen molecule process and present the antigen to T cells. B cells that have been exposed to antigen produce and secrete IgE, which binds to mast cell receptors. Memory B cells are formed from activated B cells that are specific to the antigen encountered during the primary immune response. On subsequent interaction with the antigen, cross-linking of IgE results in degranulation of mast cells. Cytokines cause chemoattraction of other inflammatory cells such as eosinophils and basophils. Neurogenic component is strongly involved in the process of onset, persistence, amplification, and extension to accompanying symptoms such as stress (Reprinted from Mandhane et al. [16]. With permission from Elsevier.)

by recent studies where allergic reactions promote the release of pro-inflammatory cytokines that directly affect the central nervous system (CNS). Ragweed has been shown to induce production of Interleukin-1 beta (IL-1 beta), one of the pro-inflammatory cytokines, in nasal epithelia that correlate with late-phase allergic responses. IL-1 beta has been shown to activate peripheral afferent nerves, which then release IL-1 beta in the brain. In turn, IL-1 beta has been shown to induce weakness, malaise, and inability to concentrate, decreased appetite, decreased activity, hypersomnia, and loss of interest in usual activities. IL-1 and IL-6 instilled intranasally in rats induced neurological dysfunction. Further, the levels of these cytokines have been found to be increased in the brains of suicide victims. Tumor necrosis factor alpha (TNF-α) that is a mast cell product also involved in IgE inflammation may also affect the CNS in a similar way (Fig. 8.3). Other pro-inflammatory mediators such as serotonin, histamine, IL-4, and IL-10 that are induced by allergies also have a direct or indirect effect on mood [16].

Another potential mechanism through which stress may induce depression in patients with allergies involves the autonomic nervous system and its effects on neurotransmitters [12]. This occurs as a result of allergy-associated overstimulation of the parasympathetic nervous system (so-called cholinergic hypersensitivity) and under-stimulation of the sympathetic nervous system (so-called beta-adrenergic hyposensitivity). It has been proposed that patients with a genetic predisposition to cholinergic hypersensitivity and who suffer from allergic reactions develop

excessive CNS cholinergic activity. This can lead to the occurrence of symptoms typically associated with depression including lethargy, feelings of being drained, slowed thinking, and social and emotional withdrawal [12].

As demonstrated in this review, there are both cellular and possibly autonomic neural influences on mood and allergies. Wright suggests that "psychological stress should be conceptualized as a social pollutant which can be 'breathed' into the body and disrupts a number of physiological pathways similar to how air pollutants and other physical toxicants may lead to increased risk of atopy [6]."

The interplay between psychological, neurological, and immunological mechanisms has also been seen in the routine treatments of allergy. Hyposensitization (immunotherapy) benefits have been documented in numerous double-blind studies, yet there is an additional placebo effect shown consistently in studies that adds further clinical benefit [17]. A striking example of this is the Rinkel method, an old form of immunotherapy using very small amounts of dilute allergen, which is now considered ineffective; yet many people improved with its use. This may have represented a placebo effect [8]. In addition, studies evaluating antihistamine efficacy in allergic rhinoconjunctivitis also have shown a significant placebo effect, which raised the question of whether antihistamines were justified in the treatment of allergic rhinitis [17]. Finally, reports that hypnosis could increase or reduce skin test sensitivity offer further evidence of the influence of psychological factors on atopy [7].

Nonallergic Rhinopathy

Nonallergic Rhinopathy (NAR) generally is given as a diagnosis to a substantial population of patients presenting to allergy practices. It has been estimated that more than 19 million individuals suffer from NAR in the USA [18]. A suspicion of NAR is raised when symptoms occur with negative allergy testing or testing that is inconsistent with relevant clinical exposure. Causes of NAR that can mimic an allergy are listed in Table 8.1.

Table 8.1 Chronic rhinitis syndromes that are nonallergic and noninfectious and not due to anatomical/mechanical causes

• Drug-induced rhinitis including rhinitis medicamentosa
• Gustatory rhinitis
• Hormonal-induced rhinitis, including the rhinitis of pregnancy
• Nonallergic rhinitis with eosinophilia
• Senile rhinitis
• Cerebral spinal fluid leak
• Nonallergic rhinopathy (NAR), previously known as vasomotor rhinitis (VMR), or idiopathic nonallergic rhinitis

Adapted from Kaliner [18]. With permission from Elsevier

AR and NAR including vasomotor rhinitis (VMR) frequently overlap and are referred to as "mixed rhinitis [19]." Because of the heterogeneous and often disparate nature of NAR, it has been suggested that these clinical conditions be classified as "Nonallergic Rhinopathy [20]".

VMR is the most commonly diagnosed NAR. It is more commonly perennial and can occasionally be associated with conjunctival and asthmatic conditions; therefore, it often is confused with AR. VMR symptoms are typically elicited by non-immunologic stimuli and environmental conditions such as cold air, changes in temperature, barometric pressure and humidity, irritants, strong odors, posture changes, and emotion [21]. The underlying mechanisms of VMR are not well understood. A hyperactive autonomic nasal reflex is suspected. Emotions and stress likely induce symptoms by their effects on the hypothalamus. In response to hypothalamic stimuli, there is an increase in the parasympathetic tone of the superior salivary nucleus resulting in relative parasympathetic predominance [22]. Other evidence suggesting a neurologic mechanism driving NAR are studies involving endoscopic resection of the vidian nerve. The vidian nerve is formed by the union of the greater and deep petrosal nerves and has autonomic fibers that are important in the balance of parasympathetic and sympathetic influences. Its resection reduces both rhinorrhea and congestion in VMR patients [19]. VMR typically occurs more in women, therefore also suggesting a hormonal role in this setting.

Psychological Influences on NAR

It has been suggested that VMR should not be regarded as a disease but as a local reaction to the environment, which varies according to patients' general physiological and psychological states prevailing at the time [23]. Rees suggested that up to 57 % of VMR episodes were precipitated by emotional factors. These included anxiety, tension, anger, hostility, humiliation, resentment, indignation, grief, and pleasurable excitement. In the same article various personality types and neurotic symptoms were reported to occur at a higher frequency in patients with VMR compared to controls, including patients who were rated as being unstable, very unstable, very meek, very anxious, cyclothymic, or hypochondriacal [24]. Conversely, Fennel found no statistical difference in psychiatric diagnoses compared to controls and no response to antipsychiatric medication in alleviating VMR symptoms [25].

Rosedale noted that neurosis frequently is associated with a stuffy nose, postnasal drip, and nasal dryness. He suggested that patients "should retain their nasal neurosis as to do without it may precipitate a worse psychic condition. The neurosis is not locally in the nose but rather the nose is the organ chosen for a general neurotic constellation. His problems are best handled by a psychiatrist [26]."

Chandler and Patterson (1984) suggested that nasal neurosis be suspected in the absence of, or minimal evidence for, IgE reactivity, and a history of visits to multiple physicians including otolaryngologists and allergists. Other characteristics suggestive of nasal neurosis include obsessional concerns with nasal symptoms, desire for nasal surgery or immunotherapy, normal nasal exam, failure to respond clinically to typical nasal medications including steroids, and no radiographic evidence of nasal polyps or sinusitis. It was suggested that the patients are frequently hostile or angry at the suggestion there is no nasal disease [27].

Treatment

Little data exist regarding the ideal intervention for those in whom there is a clear psychological association with their AR or NAR symptoms. Optimizing medical therapies based on the diagnosis is ideal and might include control of environmental triggers, cromolyn nasal sprays, oral and topical antihistamines, nasal steroids and occasionally oral steroids, occasional decongestants, leukotriene receptor antagonist, antibiotics, serum immunotherapy (SIT), or allergy shots [28, 29]. But if psychological symptoms persist, then various therapies that may be beneficial include hypnosis (Chap. 21), biofeedback (Chap. 20), or cognitive behavior therapy (Chap. 19).

NAR in Systemic Disorders

There are numerous systemic disorders that present with NAR as part of their symptom complex (Table 8.2). Some of these may involve emotional/psychogenic overlap as part of their symptom complex including fibromyalgia,

Table 8.2 Medical conditions with NAR symptoms

Metabolic
• Acromegaly
• Pregnancy
• Hypothyroidism
Autoimmune
• Sjogrens syndrome
• SLE
• Relapsing polychondritis
• Churg–Strauss Syndrome
• Wegener's granulomatosis
Other
• Cystic fibrosis
• Kartagener's or ciliary dysfunction syndromes
• Sarcoidosis
• Immunodeficiency
• Amyloidosis
• Chronic fatigue syndrome
• Gastro-esophageal reflux and laryngopharyngeal reflux

Adapted from Kaliner [18]. With permission from Elsevier

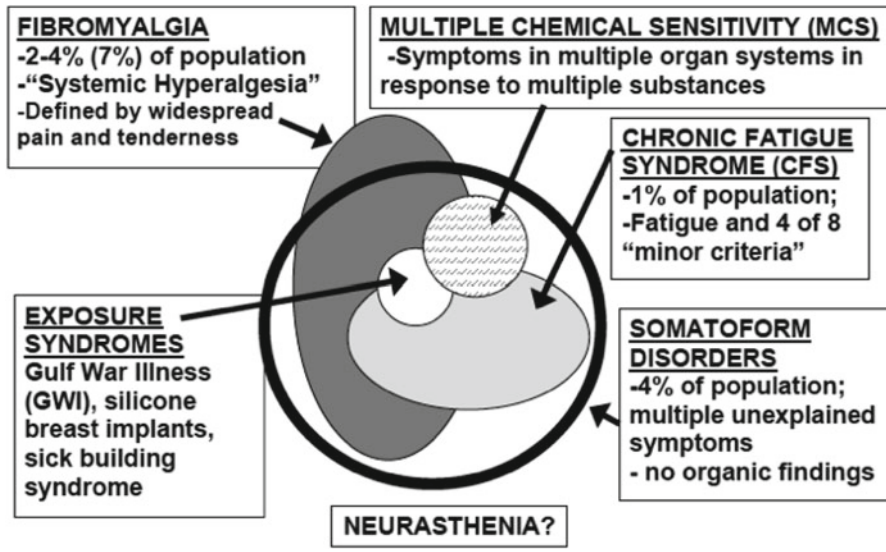

Fig. 8.4 Relationships among rhinitis, fibromyalgia, and chronic fatigue (Reprinted from Baraniuk and Zhneg [30]. With permission from Oceanside Publications.)

chronic fatigue syndrome (CFS), and multiple chemical sensitivity (MCS) (Fig. 8.4).

Fibromyalgia has a symptom complex characterized by widespread pain for at least 3 months, tenderness at least in 11/18 sites of point tenderness, and generalized aches, pains, and stiffness (Chap. 11). Usually, its symptoms are associated with exercise intolerance, poor sleep, headaches, irritable bowel, numbness, and stress or anxiety [30]. Fibromyalgia may have a prevalence rate of 2–4 % in the general population. Chronic rhinitis associated with sinus pressure, postnasal drip, and stuffiness are common symptoms in these patients. In a study from 1992, 38 % of patients who had chronic rhinitis fulfilled the criteria for fibromyalgia [31].

CFS is seen in about 1 % of the population and is characterized by unexplained, persistent fatigue for at least 6 months, impaired memory or concentration ("brain fog"), extreme malaise after activity, unrefreshing sleep, muscle pains or myalgias, headaches, multi-joint pain or arthralgias, sore throats, and tender lymph nodes. Baraniuk suggested that 2/3 of CFS patients meet the criteria for diagnosis of fibromyalgia. Compared to matched controls, patients with CFS had significantly more depression, anxiety, anger, and a lower quality of life. Typical rhinitis symptoms affect three quarters of patients with CFS, similar to the rate reported in patients with fibromyalgia, including sinus and ear congestion, mucus hypersecretion, postnasal drip, sore throat, sneezing, and runny nose, typically without nasal itch [32]. However, rates of IgE reactivity were no different in CFS patients compared to controls [33], and there was no relationship between IgE levels and nasal symptoms in these patients. Furthermore, atopy was not felt to be more prevalent in patients with CFS compared to the general population.

MCS sometimes is referred to as "20th Century disease" or "idiopathic environmental intolerances [34]." The clinical manifestations of this disorder are characterized by different systemic dysfunctions alone or in combination. These include CNS symptoms such as headache, malaise, extreme tiredness, poor memory, poor concentration, and dizziness; psychological symptoms such as moodiness characterized by irritability and anxiety; rheumatologic symptoms such as arthralgias and myalgias; GI symptoms such as bloating and diarrhea; skin symptoms including rashes; and renal or gynecological disturbances frequently attributable to food

intolerances or candida infections (leading to the hypothesized "yeast connection"). Asthma and rhinitis symptoms with stuffiness, postnasal drip, and a dry cough all are very prominent in this population [35]. Rhinitis is present in up to 91 % of these patients overall. Up to 66 % of patients complain of rhinitis after an acute "exposure" to a variety of stimulants including chemicals, strong odors, allergens, or irritants. Stuffy and full sinuses are other commonly reported symptoms.

MCS occurs most often in females (4:1), many of who are well educated, and have an extreme sensitivity (hypersomnia) to strong odors or chemicals that normally do not elicit clinical or toxic responses. Most of these patients are preoccupied by the belief that they are chemically sensitive or intoxicated with poisons. They often seek medical attention, isolate themselves socially, and have difficulty with interpersonal relationships. They adopt lifestyle changes such as wearing protective masks and altering their diet, housing, and work environment in an attempt to avoid "triggers." There are no known tests available, nor identifying clinical markers for this disorder. Diagnosis is based on clinical assessments, but since there is no generally accepted unifying theory regarding its pathogenesis, it is an extremely controversial diagnosis [36]. Most would characterize MCS as a somatoform disorder [34].

Psychogenic (Factitious) Sneezing

Sneezing is an involuntary, protective reflex typically occurring after stimulation of the nasal cavities by chemical, tactile, mechanical, infectious, or allergic triggers. Sensory nerve fibers transmit impulses to the trigeminal ganglion, and the sneezing center is reached at the lateral medulla. Upon threshold stimulation of sensory nerve fibers, the eyes close, rapid deep inspiration is followed by a forced expiration with initial closing of the glottis that leads to increased intrapulmonary pressure. Heart rate changes only minimally. The sudden dilation of the glottis gives an explosive exit of expired air through the mouth and nose. Sneeze droplets are estimated to be in the range of 40,000 droplets per sneeze, and the speed of sneeze exhalation ranges from 150 to 1,450 km/h [37].

Table 8.3 Clinical characteristics of children and adults with psychogenic sneezing

Typically occurs during adolescence
Females > males
Eyes open
Usually no nocturnal symptoms
Occur suddenly (even at rest)
No typical triggering factors
Variable duration—bouts last up to 2 h, about 30–100 per min
May be quickly regressive
Can carry on conversations, eat, and maintain normal demeanor despite sneezing
Dry and devoid of nasal secretions
An aborted sneeze or "pseudosneeze"
Physical exam and workup such as allergy test are usually negative. Refractory to a wide array of medications including even topical anesthetics
Psychiatric histories are common and triggering stressors are often identified

In 1887, Mackenzie suggested that attacks of prolonged sneezing are most likely to occur in "persons of nervous temperament [10]." The earliest published report on recurrent or intractable sneezing due to psychogenic factors was in 1949 by Shilkret. Since then there have been at least 40 more cases reported in the adult and pediatric literature. Typically, bouts of sneezing last up to 2 h, and individual sneezes occur at a frequency of 30–100 times per minute. Clinical characteristics consistent with this disorder are listed in Table 8.3 [38]. The treatment is usually successful as psychogenic sneezing responds well to hypnosis (Chap. 21), biofeedback (Chap. 20), psychotherapy (Chap. 19), or relaxation exercises. A recent publication suggested a topical local nasal anesthetic might be attempted to resolve acute symptoms [39]. Several authors have suggested haloperidol as medication alternative for intractable sneezing [40, 41].

There are several medical conditions that should not be confused with psychogenic sneezing. Many allergic and NAR symptoms can cause sneezing (Table 8.4). The photic sneeze reflex, also called the ACHOO syndrome

Table 8.4 Differential diagnosis and etiology of sneezing

Rhinitis—AR & NAR
Photic sneeze reflex (ACHOO syndrome)
Physical stimulations of the trigeminal nerve
Central nervous system pathologies
Psychogenic (Factitious or intractable) sneezing
Snatiation reflex
Sexual ideation or orgasm

Adapted from Songu and Cingi [37]. With permission from SAGE Publications

(Autosomal dominant compelling helio-opthalmic outburst), is a condition in which sneezing is triggered upon initial exposure to any bright light. Many sufferers relate that they are "allergic to the sun." Typically re-exposure has a latency period before it happens again. This syndrome is felt to be the result of a primitive neurogenic reflex that is triggered by photic stimulation of the afferent division of the ophthalmic trigeminal nerve. Sneezing can occur from pulling hair or tearing off of eyebrows as a result of mechanical stimulation of the trigeminal nerve. There are case reports of people sneezing upon sexual ideation or after orgasm. No psychiatric abnormalities are suggested. The Snatiation (sneeze and satiation) reflex is an uncontrollable sneezing attack due to stretching of the stomach as a result of overeating. This reflex has a familial pattern that may be autosomal dominant. Some CNS pathologies may be associated with sneezing such as temporal lobe or grand-mal seizures, and some patients with seizure disorders present with auras manifesting as sneezing or hiccups. Lateral medullary syndrome (LMS) often results from occlusion or dissection of the vertebral artery. The sneezing center was localized to this area of the brain as a result of the study of patients with this syndrome [37].

Food Allergy and Psychological Influences

It is important for the clinicians to recognize the different clinical presentations of food allergies before suspecting psychological issues or complications in their patients. The symptoms of food allergy are varied and unpredictable in each individual and typically mimic elements of anaphylaxis (Table 8.5). In children, hen eggs, peanuts, milk, soy, wheat, corn, tree nuts, and seed proteins account for over 90 % of reactions. In adults, allergic reactions are more likely to be triggered by fruits, vegetables, and fish. Most foods need to be ingested to cause symptoms, but rarely vapors can cause reactions. Cases of inoculation have even occurred with osculation (kissing) [42]. Specific clinical manifestations of food allergy include oropharyngeal itching and cutaneous symptoms such as hives, eczema, pruritus, and generalized rashes. GI symptoms such as nausea, vomiting, and diarrhea are common. Asthma and rhinitis do occur as a result of food allergies but are uncommon except in acute anaphylaxis. Symptoms can be delayed especially cutaneous involvement such as eczema. Severity of food allergies generally correlates with the level of food-specific IgE within the body. However, symptoms can vary based on amount of food ingested, how much is absorbed, whether the food protein is enhanced or denatured, age of the patient, presence of other atopic disorders like asthma, co-ingestion of other highly allergenic or relatively benign foods (so called dual-provoking foods), and even exercise [43]. The clinical picture is made more difficult to interpret because some patients react to foods even when testing is negative. This occurs as a result of cross reactivity of food allergens with structurally or sequentially similar proteins or even pollens or other inhalant allergens. This is described as the *Pollen-food Allergy Syndrome* or *Oral Allergy Syndrome* [44].

The diagnostic work-up of food allergies is very similar to what is done with all atopic disorders, which is to use blood tests, skin prick tests, or a combination of both. The negative predictive value of either blood or skin test is felt to be 95 %, whereas positive predictive value has a much larger variance. Therefore, since lab testing is not always consistent with the accurate diagnosis of a particular food allergy, the gold standard in allergy practices is to perform blinded challenges of an offending food [43]. The best method is double-blind, placebo-controlled challenges wherein the patient and provider of the tests are both blinded to the content of the administered

Table 8.5 Food allergies and their clinical manifestations

Target organ	Immediate symptoms	Delayed symptoms
Cutaneous	Erythema	Erythema
	Prutitus	Flushing
	Urticaria	Pruritus
	Morbilliform eruption	Morbilliform eruption
	Angioedema	Angioedema
		Eczematous rash
Ocular	Pruritus	Pruritus
	Conjunctival erythema	Conjunctival erythema
	Tearing	Tearing
	Periorbital edema	Periorbital edema
Upper respiratory	Nasal congestion	
	Pruritus	
	Rhinorrhea	
	Sneezing	
	Laryngeal edema	
	Hoarseness	
	Dry staceato cough	
Lower respiratory	Cough	Cough, dyspnea, and wheezing
	Chest tightness	
	Dyspnea	
	Wheezing	
	Intercostal retractions	
	Accessory muscle use	
Gastrointestinal (oral)	Angioedema of lips,	
	Tongue, or palate	
	Oral pruritus	
	Tongue swelling	
Gastrointestinal (lower)	Nausea	Nausea
	Colicky abdominal pain	Abdominal pain
	Reflux	Reflux
	Vomiting	Vomiting
	Diarrhea	Diarrhea
		Hematochezia
		Irritability and food refusal with weight loss (young children)
Cardiovascular	Tachycardia (occasionally Bradycardia in anaphylaxis)	
	Hypotension	
	Dizziness	
	Fainting	
	Loss of consciousness	
Miscellaneous	Uterine contractions	
	Sense of “impending doom”	

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food or placebo capsules, in order to remove potential bias of the physician's own impressions, and the patient's subjective reactions [45].

Many clinicians prefer single-blind challenge as it is more practical to blind only the patient. An open challenge in which the patient knows which food is being given is the least preferred. This is sometimes performed when it is difficult to hide the taste or smell of a food, or when the patient is very young. Food challenges can be time consuming, difficult to prepare, and frequently depend on observation and subjective complaints as their major assessment tool.

The importance of accurately diagnosing food allergies cannot be underscored enough. A diagnosis of food allergy can impose a psychological burden on patients and parents, affect quality of life, require anaphylaxis education and prevention, and in many situations may place an undue restrictive nutritional burden, especially on children [46]. For example, of 11 patients who developed failure to thrive because of their parents' beliefs about allergies to multiple foods, only two reacted to a single food by oral challenge [47].

Parents of children who are allergic to peanuts have to deal with considerable disruption in their child's daily activities and impairment in family/social interactions. Further, in comparison with parents of children with rheumatologic disorders and diabetes, parents of children with peanut allergy have been shown to increase rates of anxiety related to their children's increased risk of death [48].

The disruption to life activities as a result of food allergies may be the reason adolescents might engage in undue risk taking in relationship to food, such as a result of not reading labels carefully or not carrying epinephrine when a true food allergy exist. Further compounding the issue for teens are their reduced appreciation of potential life-threatening dangers, a belief that consequences can be controlled, and a fear of social isolation [49].

Because of the difficulties in diagnosing food allergies accurately, the epidemiology of food allergies is poorly understood. A recent large-scale meta-analysis of the prevalence of food allergy suggested that the prevalence of self-reported food allergies was 12–13 %. However, when subjected to rigorous scientific verification the overall prevalence was much lower [50]. Children have a higher incidence of true food allergy, which affects 6–8 % of all children and has been increasing in incidence over the past 2 decades. In contrast, food allergy affects only 1–4 % of adults, and the incidence rate has remained stable [51].

There are no easy explanations for the increasing incidence of food allergies in infants but the "hygiene hypothesis" has been postulated. Broadly stated, this hypothesis suggests that natural infections and microbial exposure protect against allergy. Thus, the recent trends toward rising atopic disorders including food allergies may be as result of decreased natural infections due to vaccinations, overuse of antimicrobials, and the urbanization of society away from rural and farming living [52].

Perceived food allergy is an extremely common complaint and most studies suggest that food intolerance accounts for 90 % of all symptoms attributed to food allergy. Food intolerance occurs for a variety of reasons without a demonstrable immunologic basis and tends to be a catchall diagnosis. Frequent clinical complaints generally center on the gastrointestinal tract such as altered bowel pattern or recurrent abdominal pain. Because the pathways and mechanisms underlying food intolerance are not always clear, symptoms are frequently classified as "functional" without defining the real cause of the problem.

Just like with rhinitis, in which entopy has been demonstrated, a local allergic reaction may exist despite a lack of systemic food-specific IgE [53].

Lied found that up to 61 % of patients with self-perceived food disorders had other atopic phenomena such as asthma, AR, and eczema. These patients had a higher numbers of IgE positive mast cells and increased intestinal permeability as compared to controls [54].

There are other disorders such as gluten sensitivity, eosinophilic esophagitis (EE), and allergic colitis that have complex immune or mixed mechanisms that may or may not include IgE as part of their mechanism. These conditions have

slightly different clinical presentations as compared to problems induced by true food allergy. For instance, EE affects all ages and clinically can present with choking on foods (dysphagia), impaction, vomiting, regurgitation, and feeding disorders [55, 56].

Celiac disease and now gluten sensitivity are considered the expression of different forms and severity of an immune-mediated sensitivity to gluten, a protein seen in common grains such as wheat. These conditions are frequently confused with wheat allergy, but are not IgE mediated. These conditions present with gastrointestinal symptoms but rarely have been associated with cutaneous or respiratory symptoms as seen in true wheat allergy. Instead, these symptoms are associated with anti-endomysial, anti-tissue transglutaminase, and anti-gliadin antibodies [57, 58].

The term "pseudo-food allergy" has been used to describe people who falsely believe that they have symptoms attributable to food allergies [59]. Many studies have confirmed that patients suspect food hypersensitivity is a cause of their clinical problems, even when true allergy cannot be documented. For example, in a recent study of 200 children with recurrent abdominal pain of at least 3 months duration, only 5 (2.5 %) had a food allergy documented by double-blind placebo oral challenge, despite a "suspicion" of a food allergy in 123 (62 %) of the patients. Abdominal pain due to a functional disorder was directly attributable to stress in 2.5 % of these children. The majority of the children in this study were diagnosed with unrecognized constipation or their symptoms resolved spontaneously [60].

Affective and other psychological conditions with somatic complaints, which could be interpreted as features of anxiety or depression, are prominent in patients in whom food hypersensitivity cannot be confirmed [45, 61]. Early reports have suggested a putative association of food allergies and symptoms of attention deficit hyperactivity disorder, but clinical studies have generally repudiated any association [62].

Pearson found a high incidence of psychiatric disorders such as anxiety and depression, as well as high suggestibility, in adults who believed they had a food allergy. This group was identical in psychiatric symptomatology compared to a group of psychiatric outpatient referrals [61]. Another study from Norway found that of patients with self-reported food hypersensitivity, 57 % fulfilled the DSM-IV criteria for at least one psychiatric disorder including anxiety (which affected 34 %) and depression (16 %) [63]. Vatn found that 13 of 17 patients with Non-IgE-mediated food hypersensitivity had major life stressors such as major distress or trauma in childhood including loss of a parent, violence, or major psychiatric disorder [64]. Another study comparing non-IgE-mediated food hypersensitivity with normal volunteers and healthcare workers found significantly more subjective health complaints such as tiredness, bloating, and headaches in the hypersensitive group [65].

On the other hand, earlier studies suggested food intolerance is not associated with psychiatric disorders. Peveler et al. found in a community study of 273 patients who complained of food intolerance and not judged to be clinically allergic, there was no mood disturbance, impaired social adjustment, or other psychological symptoms. A limitation of this study was that this cohort of patients represented a population with mild symptoms as they did not seek specialist care [66]. Rix studied 23 patients in an allergy clinic and found no significant psychiatric disorders associated with food intolerance. Giving placebo of a suspected food did not induce a psychiatric or psychologically consistent response [67].

In rare cases foods may induce psychotic symptoms. Denman described a case of a 14-year-old girl who developed "mental disturbance" upon milk exposure, which included irrational bouts of crying, hysteria, withdrawal, hearing voices, and illusions. These episodes were provoked by double-blinded administration of milk and blocked by the mast cell-stabilizing agent sodium cromoglycate [68].

Psychosomatic Food Allergies

Cases of psychosomatic food allergy have been reported rarely. As in many other disorders, a clinical history is vital, as true IgE-mediated

disease has distinctive clinical characteristics. A true food allergy typically presents with hives, oropharyngeal itching, generalized red rash or flushing, and vomiting. Asthma and/or rhinitis symptoms can occur. A delayed reaction such as eczema can develop. Headaches, dizziness, irritability, hyperactivity, somnolence, fatigue, and poor concentration are unlikely to be attributable to a true food allergy [69]. Other gastrointestinal diseases that may mimic allergies or have an allergic overlap such as gastro-esophageal reflux (GERD), EE, or gluten sensitivity present with mostly GI symptoms. Vomiting alone can occur such as is seen in fish allergies but usually most allergic reactions start within minutes and no later than 2 h after ingestion of a food.

Minimal sensitivity to foods by allergy testing is documented commonly, and in such instances, food challenges are necessary to rule out clinically significant reactions. Having asthma and allergies which may be seen commonly in food allergic patients does not preclude the possibility that psychosomatic food associated symptoms can exist.

In 1954, Kaufman, a practicing generalist, wrote a comprehensive diary/observation of about 600 patients who had some form of "idiosyncratic food reactions." According to Kaufman:

> *Each individual very early in life has developed conditioned responses to eating and to certain foods. His conditioned patterns of reacting are the result of his individual integration of behavior learned from his culture, his religion, parents and from his own life experiences…Allergic reactions to foods tend to evoke a postnoxious behavior pattern which becomes a significant part of the total reaction to the offending food…. The foods tend to be omitted from the individual's diet* [70].

Kaufman found that although some patients continued to eat foods to which they may have been mildly allergic, the majority reacted adversely and avoided foods that triggered their allergic reactions. Further, he identified many patients who did not have food sensitivities but continued to report symptoms, perhaps in order to gain concessions from family, friends, and employers (secondary gain). He referred to such behavior as a "cleaning-up" or "setting-into-order" reaction, in which patients try to arrange aspects of their physical environments, work situation, or emotional reactions into an orderly scheme. This pattern of conditioning might also explain why some patients continued to avoid certain foods even if they outgrew their allergy.

Another aspect of conditioning that may occur is associated with smell or odor. Allergic reactions can occur when food proteins are cooked and convert from a solid phase to a vapor phase. Fish and shellfish are classic examples. Other examples include peanut dust being disseminated from the husking of peanuts in a closed environment. Most of these events will lead to ocular itching and sneezing, while generalized anaphylaxis would be rare.

There are a number of anecdotal reports of allergic reactions to the smell of peanuts or peanut butter. To clarify this, Simonte et al. studied 30 highly sensitized children with peanut allergy and subjected them to a concoction of peanut butter masked with tuna and mint; soy butter was used as the placebo control. In addition, peanut or soy butter was placed on the skin to see if casual contact elicited systemic clinical responses. There were no clinical respiratory systemic reactions to either inhalation or contact of peanut butter [71]. The explanation for odor-related allergic reactions most commonly put forth is based on the concept of classical conditioning (also known as a Pavlovian response). In this situation, patients learn to associate the smell of a food with the allergic physical reaction elicited by a past ingestion of the food. Once conditioning occurs, patients develop the allergic reaction upon exposure to the smell alone. Almost any physiological response can become conditioned ranging from rapid heart rate, flushing, itching, hives, wheezing, and even high blood pressure. In addition, conditioning may also explain the development of panic or fear that may be confused with a severe allergic reaction with associated symptoms such as increased heart rate, chest tightness, shortness of breath, and difficulty talking. Panic attacks can mimic allergies and clinically be difficult to differentiate [72].

Treatment

Little data exist regarding the ideal intervention for those in whom there is a clear psychological association with food allergies or intolerance. Various therapies may be beneficial including hypnosis (Chap. 21), biofeedback (Chap. 20), or cognitive behavior therapy (Chap. 19).

Case Studies

Case 1: Sneezing

A 17-year-old African-American female in her junior year of high school was sent to the school nurse for disrupting her class in the middle of the school year. She presented with loud, obnoxious, continuous sneezing. The school nurse obtained a brief history. She had known the patient because of her history of asthma and seasonal allergies. She was a good student but struggling with grades this year. However, her asthma was under good control. Previous allergy tests were positive for pollen only.

The patient stated she had developed a mild upper respiratory infection the previous week and yesterday woke up with sneezing, nasal congestion, but no rhinorrhea. She had no sneezing throughout the night. She tried her oral antihistamine (cetirizine) and nasal steroid without relief. The nurse noted the student was sneezing with her eyes wide open and was sneezing up to 50 times per minute. She was referred to her allergist later that day and the sneezing did stop during lunch but recurred in the allergist's office where it was heard throughout the waiting room. The patient was sneezing and could deliver a history while she was sneezing. The physical exam was benign with no postnasal drip nor secretions noted. Her turbinates were normal in color and minimal bogginess was noted.

Questions

1. What is the patient's likeliest diagnosis?
   (a) Vasomotor Rhinitis
   (b) Upper respiratory infection
   (c) Factitious sneezing
   (d) Allergic Rhinitis
   (e) Solar Rhinitis
2. Psychogenic sneezing is characterized by all EXCEPT:
   (a) Paucity of nasal secretions
   (b) Sneezing only while awake
   (c) Sneezing with eyes open
   (d) More common in females
   (e) Sneezing with sun exposure
3. What is this patient most likely benefit from as initial therapy?
   (a) Haloperidol
   (b) Valium
   (c) Allergy Skin test
   (d) Sinus imaging study
   (e) Biofeedback or hypnosis

Answers

1. (c): This case would typify a classic presentation of psychogenic sneezing. It is more frequently seen in adolescent females; asthma and allergies can be present but her particular pollen allergy was not relevant during wintertime in the middle of the school year. This patient was a good student but was under stress related to her academic performance in her junior year. The frequent sneezing, the normal physical exam, the disappearance at night, and her easy distractibility during lunch make the diagnosis very likely. Response to medications such as antihistamines or nasal sprays is transient at best.
2. (e): Sneezing with sun exposure is called Solar Rhinitis and is a primitive neurogenic response to photon ultraviolet light stimulation. This is a temporary condition, which resolves quickly. Psychogenic sneezing typically has very little secretions expelled, patients frequently sneeze with their eyes open and shut, and resolves with sleep or being easily distracted.
3. (e): Medications such as Haldol or Valium can be used in intractable, unresponsive situations. But there are not appropriate as an initial response to treatment. Psychogenic sneezing responds well to hypnosis (Chap. 21), biofeedback (Chap. 20), cognitive behavior therapy (Chap. 19), or relaxation exercises.

Case 2: Food vs. Psychosomatic Allergy

A 16-year-old female with asthma and allergic rhinitis developed generalized hives, shortness of breath, and wheezing, 20 min after ingesting peanut butter. She went to the emergency department where it was noted she had scratch marks over her body. The next day she had a similar reaction after ingesting a candy with peanut butter. Subsequent evaluations by blood and skin test were negative. An open challenge with a teaspoon of peanut butter induced flushing, hives, and linear welts, but no respiratory signs. The patient then underwent a double-blinded, placebo-controlled challenge in which she wore nose clips to decrease her sense of taste and smell, and the peanuts were chopped and mixed with pecans, cinnamon, and walnuts to conceal their taste. She had four challenges and the reactions were never reproduced. Later, with open administration of peanuts and peanut butter, the reactions never recurred [73].

Questions

1. The best way to differentiate a true food allergy from a psychogenic food allergy is:
   (a) Skin Prick Test
   (b) CAP/RAST blood test
   (c) Single-blind food challenge
   (d) Double-blind placebo-controlled challenge
   (e) Patch test
2. Foods can induce allergic reactions typically by all of the following mechanisms *EXCEPT*:
   (a) Smelling
   (b) Kissing
   (c) Ingesting
   (d) Cross-contamination
   (e) Intradermal skin testing
3. All of the following symptoms typically are associated with IgE food-mediated allergy EXCEPT:
   (a) Oral-pharyngeal itching
   (b) Hyperactivity
   (c) Hives
   (d) Eczema
   (e) Cough

Answers

1. (d): Although all the above tests (skin prick, CAP/Rast, Patch) may be valuable in screening and initial evaluations for food sensitivity, they all suffer from significant false positive and false negative predictive values. Single-blind challenges still may suffer from administrator bias; therefore, the gold standard of diagnosing a food allergy is double-blind placebo-controlled food trials.
2. (a): Most food allergies and subsequent clinical reactions occur directly with ingestion or direct inoculation into the body. Kissing and exchanging saliva would be a potential mechanism. Cross-contamination occurs when a food allergen is mixed inadvertently with another seemingly innocuous food. Intradermal skin testing that are used for inhalant allergy testing is not recommended for foods as the risk of introducing an allergen into the blood stream is high and therefore there is a higher risk of systemic reactions. Simple contact or smelling would rarely cause a reaction.
3. (b): Hyperactivity and behavioral abnormalities have been suspected for years to be associated with food allergies, but studies have never proven causality. Any typical IgE-mediated reaction typically involves the skin, respiratory tract, or GI system. Although immediate reactions predominate, delayed reactions such as eczema may occur. Oropharyngeal itching is a very common symptom especially in milder allergies that occur with fruits and vegetables.

Conclusions

Psychological influences on allergies such as related to food and AR have an important impact on disease diagnosis and management. In many cases, understanding the differences and interactions between physiological and psychological aspects of patient presentation are vital. Such an understanding may not only be life saving but also can direct the proper management of the disease state and may even lead to ultimate resolution of symptoms.

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Functional and Psychosocial Ramifications of Type 1 Diabetes Mellitus in Pediatric Endocrinology

9

Nicole L. Pilek and Harold S. Starkman

Abstract

Type 1 diabetes mellitus is a common chronic medical disorder of childhood and adolescence that requires intensive day to day management. In addition to measuring dietary intake, children and adolescents with diabetes must test their blood sugar and administer insulin multiple times daily. They are also responsible for correcting high and low sugars. A comprehensive diabetes knowledge base and effective family communication and problem solving skills are necessary to optimize blood sugar control, so as to avoid both short- and long-term diabetes complications. Diabetes management requires a unique therapeutic alliance in which patients and their families manage diabetes, and their healthcare providers serve as mentors and consultants.

Although diabetes-related stresses in themselves can be overwhelming, these stresses are often complicated by patient-related psychosocial stressors such as anxiety, an eating disorder, or depression. In addition, family and larger system challenges such as marital discord and the presence of other family chronic disease or financial pressures may also interfere with optimal diabetes management. This can result in adherence issues, functional symptoms, or suboptimal diabetes control. To complicate matters further, psychosocial issues often can lead to suboptimal blood sugar control, which in itself, can cause psychosocial distress.

Since blood sugar control and psychosocial factors are so closely linked, it is imperative that a whole person approach be followed for optimal diabetes management. Diabetes issues, especially those related to

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regimen adherence, need to be evaluated from a psychosocial as well as medical perspective and interdisciplinary solutions should be offered. In addition, research suggests that successful diabetes management requires a collaborative approach by medical and mental health professionals familiar with type 1 diabetes and its treatment.

Keywords

Diabetes mellitus • Type 1 diabetes • Childhood diabetes • Depression • Eating disorder • Blood sugar • Anxiety • Adherence

Overview

Type 1 diabetes mellitus is the second most common chronic medical disorder of childhood and adolescence after asthma, with a reported prevalence of 1:500 in the USA [1]. Type 1 diabetes incidence has been increasing at a reported rate of 3 % annually with a shift toward increased incidence in children under the age of 5 years [2]. The disorder is characterized by autoimmune destruction of insulin producing islet cells of the pancreas, leading to insulin deficiency, which, in turn, prevents sugar from entering cells where it can be utilized as fuel. There is only a weak genetic predisposition to the development of type 1 diabetes in youth. This is reflected in the fact that over 90 % of youth diagnosed with this disorder have no history of type 1 diabetes in their immediate or extended family [3].

At the time of diabetes onset, symptoms often include increased drinking and urination, as well as bedwetting. Left untreated, elevated blood sugar can produce urinary fluid losses that exceed oral intake, especially in younger children. This can lead to a severe life threatening form of dehydration known as diabetic ketoacidosis.

Effective diabetes management includes balancing food, medication, and activity with multiple daily finger stick blood sugar measurements. The goal of therapy is to optimize blood sugars while minimizing significant hypoglycemia and allowing the child or adolescent to live as normal a life as possible. This approach has been shown to minimize risk for the development of long-term, diabetes-related eye, kidney, and nerve complications [4].

Medical management of type 1 diabetes includes insulin replacement, either by multiple daily injections or by an insulin pump, a device worn by the patient, which delivers a continuous flow of rapid acting insulin through a subcutaneous catheter. For both regimens the child or his/her parents must weigh meals and snacks to calculate carbohydrate content, as well as perform finger stick blood sugar testing multiple times daily. This information is then utilized to determine insulin doses, which are administered by injection or pump at each meal or snack. Physical activity is encouraged, but must be integrated into the diabetes treatment plan. The complexity of diabetes management requires intensive continuing family diabetes education and support from the time of diagnosis. Current management guidelines recommend that education, as well as medical and social support for patients and their parents be provided by a multidisciplinary team composed of a pediatric endocrinologist, certified diabetes nurse educator, dietitian, and mental health professional [5].

Since most diabetes management occurs at home, away from the clinic or hospital, families are primarily responsible for disease management. Home management of diabetes requires a significant diabetes knowledge base, as well as effective family communication and problem-solving skills. To complicate matters further, effective diabetes management frequently is impacted by and can generate additional day-to-day family stresses, often exacerbated by developmental, family, and other systemic psychosocial influences. These stresses necessitate a comprehensive "whole family" management approach, which merges medical, educational, and psychosocial support.

Psychosocial Considerations[1]

Effectively managing diabetes continually influences all aspects of a young person's day to day life, an effect that often reverberates through the whole family. All meals and snacks must be measured, evaluated for carbohydrate content, and then covered with appropriate insulin doses. Blood sugars need to be measured multiple times each day and out-of-range sugars treated. Long-term blood sugar patterns need to be reviewed by the patient and parent, and regimen changes made. These diabetes-related "chores" are superimposed on the child or adolescent's baseline school, home, and other responsibilities.

Associated with responsibilities related to diabetes management are the emotional concomitants of having and caring for an unwelcome and demanding medical disorder. Often youth with diabetes feel different from their peers, a feeling exacerbated by visits to the school nurse and a medical routine involving needles and blood, which can be discomforting for all involved. In addition, the constant fear of having a low blood sugar reaction, especially in the presence of friends, can lead to diabetes mismanagement to keep blood sugars "safely" elevated. For young patients, the challenges related to having diabetes can overshadow those of managing common social issues such as mastering the developmental tasks associated with puberty and identity formation, changes in body image, peer pressure, and attaining independence from parents. Worry about possible long-term complications is yet another diabetes-related stress, one which often affects parents as well as their children.

In view of these stresses, it comes as no surprise that anxiety and depression are reported to be common in youth with diabetes [6, 7], as well as their parents [8]. In addition, the specific focus on diet, necessary for optimal diabetes management, makes disordered eating a problem not uncommon in adolescent girls with diabetes, with a reported prevalence as high as 14 % [9]. Diabetes regimen adherence issues are also common in youth, especially during adolescence, when developmental needs for independence and experimentation often collide with the demands of diabetes management.

Diabetes-related stress is particularly common at diabetes diagnosis. This is the time when children and adolescents are faced with adjusting to a new "normal" lifestyle and often to a changed view of him or herself. In addition, since diabetes is a condition where disease management is primarily the patient's and family's responsibility, a unique family/medical provider relationship must be forged early on, one wherein the family members serve as primary diabetes caretakers and their medical professionals as educators and consultants. These factors together with the complexity of diabetes management and influence of both psychological and external social factors can also increase stress, sometimes to levels where it becomes overwhelming. This affects quality of life for both the individual and family [10].

In addition to diabetes leading to increased family stress, both diabetes- and non-diabetes-related stresses can affect diabetes regimen adherence, resulting in poor blood sugar control and multiple diabetes-related emergency department and hospital admissions [11–14]. Such stressors commonly include depression, anxiety, adjustment issues, family dysfunction, as well as chronic parental health problems [9]. These underlying psychosocial issues are often hidden, but can be reflected in a child or adolescent's poor blood sugar control, which is seemingly unresponsive to regimen modification. Other clues of underlying psychosocial problems may include family conflict at medical visits or more overt regimen adherence issues such as the skipping of blood sugar testing and/or insulin doses. Management of these patients should include early comprehensive psychosocial evaluation by a social worker or other mental health professional.

[1] Although type 1 diabetes differs from other forms of diabetes including diabetes related to cystic fibrosis and type 2 diabetes, psychosocial stressors are common to both.

Representative Clinical Cases and Discussion

Pediatric/Adolescent Psychosocial Factors Complicating Diabetes Management

Depression

Mary, a 15-year-old with type 1 diabetes since age 5 has been hospitalized three times in the past year for elevated blood sugar and severe dehydration. Poor blood sugar control has also been documented at outpatient medical visits where her Hemoglobin A1C, (a blood test measuring average blood sugar) was elevated. Mary complains of feeling lethargic and is having trouble staying awake in school because she is up several times each night going to the bathroom. She is falling behind in her schoolwork and her grades are starting to suffer. Her endocrinologist notes that the patient has a "flat affect."

Depression is a serious but common health issue, affecting up to 25 % of children and adolescents with diabetes [15]. Depression can adversely affect quality of life and overall functioning and is associated with increased suicide risk. There are additional consequences for youth with diabetes, where depression is commonly both a cause and effect of poor regimen adherence. Chronic elevation of blood sugar, due to poor regimen adherence, can increase both diabetes-related and depressive symptoms [16]. In addition, elevated blood sugar can increase risk for long-term diabetes complications such as eye and kidney disease.

It is important to remember that symptoms of depression can be confused with those associated with uncontrolled diabetes. Feeling lethargic, having difficulty concentrating, changes in sleeping habits, poor school performance, and irritability are common symptoms of both depression and poorly controlled diabetes [17]. For this reason, depression screening of adolescents who appear to be depressed and/or have difficulties with regimen adherence is indicated when either of these findings is observed at a routine clinic visit.

Anxiety

Larry, a 10-year-old with a 2-month history of type 1 diabetes, is seen in clinic for routine diabetes follow-up. During the visit, the patient's mother tells his diabetes nurse that although Larry had previously loved playing soccer, this season he has been complaining of headaches that forced him to miss most practices and games. She questions whether Larry's headaches are somehow related to his diabetes, or if a referral to a pediatric neurologist might be indicated.

Larry's nurse encourages Larry and his mother meet with the team social worker before deciding on the need for a neurological referral. During his visit with the social worker, Larry reveals that he is "very nervous" about having his teammates see him check his blood sugar and is afraid of having an embarrassing low blood sugar reaction on the soccer field.

Anxiety-induced somatic symptoms including headache often resemble symptoms associated with high or low blood sugar. This can create a challenge when making management decisions, unless blood sugar results are available from the time symptoms were present. In this patient, blood sugar results were in the target range when his headaches occurred, making a diagnosis of anxiety-related symptoms most likely. Intervention included short-term counseling and support, resulting in headache resolution after which Larry returned to playing soccer.

Discomfort related to sharing information about newly diagnosed diabetes with friends, feeling "defective," and fear of being treated differently because of diabetes may produce somatic symptoms such as headache or abdominal pain in children and adolescents [18, 19]. It is important to keep in mind that a child complaining of symptoms that can be associated with diabetes, may, in fact be struggling emotionally, which may be expressed through physical symptoms. Often these youth drop out of activities to avoid bringing attention to their diabetes. Referral for support and counseling, after reviewing blood sugar logs to rule out diabetes-related symptoms, is usually an effective approach to resolving such functional symptoms.

Developmental Issues

Sue, a 14-year-old with a 2-year history of diabetes, has her annual dietitian visit. Her Hemoglobin A1C has been increasing steadily over the past year. Sue denies skipping insulin. She suggests that "hormones" and "guessing" the amount of carbohydrates she eats, when out with her friends, are contributing to her higher blood sugars. Upon further exploration, the dietitian discovers that Sue also does not routinely check her blood sugar or give herself insulin when she is with friends.

At a time when developing autonomy and independence become an important part of the normal developmental process, feelings of invincibility often lead to risk-taking behaviors, especially involving diabetes management [18]. In order to appear as normal as possible and often feeling embarrassed and different from friends, teens with diabetes may "forget" to check their blood sugars and fail to give insulin before they eat. Since adolescents often tend to live only in the "here and now," teens with diabetes may not consider long-term complications of their disease and seek the more easily attainable short-term goals of being like their friends and living "normal" lives.

As adolescents begin to become autonomous and explore their independence, they are expected to take on more of their own diabetes care, with less parental involvement. Successfully making this transition requires a delicate balance of parental supervision and "letting go." Establishing a safe place for parents and adolescents to work out these issues is an essential function of the diabetes care provider.

Eating Disorder/Disordered Eating

Cynthia, a 17-year-old girl with type 1 diabetes of 4 years duration, is seen in clinic for routine diabetes follow-up. Since her last visit, 3 months previously, Cynthia's weight has decreased by 8 pounds and her Hemoglobin A1C has increased to 10%, indicating that her blood sugars have been significantly elevated. Although Cynthia "forgot" to bring her blood sugar results, she says that she has been testing four times daily, as recommended, and that most of her sugars are in the normal range. Cynthia does reveal that her junior prom is next month and that she is nervous about looking good in her new dress. When asked specifically about missed insulin doses, Cynthia admits that she "occasionally" skips insulin before meals and snacks, especially when she needs to lose weight.

Studies have shown that up to 16 % of adolescent girls with diabetes omit or reduce their insulin doses, as a means to lose weight [9, 20]. A desire to be thin and viewing body shape and weight as measures of self-worth are common issues that adolescent girls must navigate. In a small proportion of these girls, weight concerns result in the development of eating disorders, characterized by self-induced vomiting, overexercising, and/or the use of diuretics and cathartics. In adolescents with diabetes, symptoms of an eating disorder may also include under dosing or omitting insulin. Skipping pre-meal insulin prevents dietary sugar from entering cells where it can be utilized as fuel. This results in elevation of blood sugar, symptoms of uncontrolled diabetes and risk for life threatening dehydration (ketoacidosis) acutely, and increased risk for diabetes-related complications over the long term.

A pattern of multiple diabetes-related hospitalizations or unexplained poorly controlled blood sugars, especially in an adolescent girl, should be an indication for eating disorder screening. Referral to a specialized eating disorders program, which closely collaborates with the patient's diabetes care team, is crucial for the successful management of these patients.

In addition to manipulating insulin to lose weight, children and adolescents with diabetes of both sexes may exhibit disordered eating behaviors or patterns. Management of diabetes requires close scrutiny of nutrition and measurement of food consumption. It also requires insulin administration before each carbohydrate-containing meal and snack. Often, diabetic children and adolescents may adopt unhealthy dietary habits such as consuming less carbohydrate and more protein than necessary, or skipping meals and snacks altogether, in order to avoid giving injections. Reducing food intake may also lead to sneaking

food, "closet eating," and possible binge eating; situations where necessary insulin dosages are usually not given. The causes underlying such dietary behaviors must be explored, since anxiety, needle phobia, or depression may be underlying factors.

Family Psychosocial Factors Complicating Diabetes Management

Parental Anxiety/Depression

A mother brings her 6-year-old son for a visit with his pediatric endocrinologist. Ricky has had diabetes for 2 years. His finger stick Hemoglobin A1C is 9.2%, indicating that Ricky's diabetes is not well controlled. Review of his home blood sugars show that Ricky has been having blood glucose checks performed 10–15 times daily and that his sugars are consistently elevated during the evening and overnight. Ricky's mother states that, since diagnosis, she doesn't get very much sleep. Since she is awake, tests Ricky's blood sugar often to make sure that he is not too low.

It is not uncommon for parents of youth with diabetes to have a fear of low blood sugars [21]. Although an occasional mild hypoglycemic reaction, easily treated with fruit juice, is a common result of targeting near normal blood sugar, a small number of children and adolescents can be unaware that their blood sugar is too low. These children may be at risk for a severe low blood sugar reaction, often requiring an emergency home injection of glucagon, or a trip to the hospital. The fear of such reactions can add stress to an already demanding regimen. Often sleep patterns are interrupted. Parents worry during the night and check their child repeatedly. In addition, fear of their child having a severe low sugar may cause some parents to maintain blood sugars at higher levels than recommended or treat borderline blood sugars prematurely. For these families, the short-term goal of avoiding an uncommon diabetes complication overshadows the longer term risk of developing diabetes complications.

Fear of low blood sugar is not the only aspect of diabetes care that can be stressful. Diabetes is a chronic disorder which requires that the patient and family assume multiple daily responsibilities related to disease management. These responsibilities can overwhelm parents and force them to question their ability to care for their child. Feelings of guilt and sadness may also affect how parents cope emotionally with their child's diabetes [22]. In addition, managing life changes associated with having diabetes, including loss of normalcy, can be enormously stressful.

Parental stressors related to diabetes, if not addressed, can lead to more serious emotional responses including marital conflict, anxiety, and depression [23, 24]. These responses can impact the emotional well-being of the whole family and affect the home medical management of the affected child or adolescent.

Family Functioning

Michael, an 11-year-old with a 3-year history of diabetes, has had four hospital admissions related to poor blood sugar control over the past 6 months. Up until this year, Michael has had excellent blood sugar control. During his most recent hospitalization, Michael's parents have told the physician that things have been difficult at home recently since they have separated and are planning to divorce.

Optimal family functioning is a predictor of therapeutic success for children and adolescents with chronic medical illness and especially for those with diabetes [8]. In pediatric patients, it has been well documented that effective family communication, family cohesion, and mutual support play an integral role in insuring adherence to the diabetes regimen [10, 25]. Studies have also shown that positive family unity, a collaborative approach to diabetes management, and mutual supportive efforts are associated with better blood sugar control [26]. In contradistinction, neglected diabetes responsibilities related to disagreements about how the diabetes is to be managed are associated with poorer blood sugar control [27].

Acute and chronic family stressors including marital conflict, severe family illness, and major life changes such as divorce or loss of employment can negatively influence parents' ability to adequately provide the attention required to

successfully manage diabetes. In addition, the child or teen, made anxious by family stress, often has difficulty focusing on his or her diabetes.

When the pediatric or adolescent patient with diabetes is not doing well, it is imperative to ask about family stresses and their possible effect on diabetes management. When indicated, referral to a skilled family therapist is an effective tool for resolving conflict as well as for improving family communication and diabetes management.

Differentiation of Functional from Organic Symptoms in Type 1 Diabetes

When things are not going well for a youth with diabetes, it is important to take a "whole person" approach and consider both medical and psychosocial factors which may affect symptoms and/or blood sugar control. This often necessitates the combined expertise of a collaborative multidisciplinary team of medical and mental health professionals, experienced in diabetes management.

The usual approach to managing these often complex patients is to rule out medical causation first. Once medical causes such as incorrect insulin dosage are ruled out, evaluation of the youth and his caretakers by a social worker or psychologist is indicated. In addition to discussing diabetes-related issues, this assessment should include a comprehensive family evaluation as well as depression and eating disorder screening, if indicated. Also important, is an assessment of the family's diabetes knowledge base and problem-solving skills. This should include direct observation of these skills, if regimen adherence issues are suspected.

Effective psychosocial intervention may include family therapy, individual counseling, cognitive behavioral therapy (Chap. 19), and problem-focused therapy [28–33]. Here too, close collaboration and communication between medical and mental health team members is critical for clinical success. Information on treatment strategies, as well as more specific information on various modes of intervention is discussed in the second section of this book.

Additional Case Studies with Self-Evaluation Questions

Case 1: Diabetes and Abdominal Pain

Since the beginning of the school year, Brian, a 7-year-old with a 2-year history of diabetes, visits the school nurse daily, just after arriving at school, complaining of "stomach pain." Brian has no other symptoms and has been eating well. At each visit, Brian's blood sugar is measured and is always in the normal range. After 45 min with the nurse, Brian's abdominal pain resolves, and he returns to class. Both the school and Brian's mother are concerned about the stomach pain, as well as about the amount of math class he has been missing.

Questions

1. The most likely cause of Brian's abdominal pain is:
   (a) Celiac disease
   (b) High blood sugar
   (c) Low blood sugar
   (d) Constipation
   (e) Anxiety
2. When evaluating the cause of Brian's abdominal pain, it would be least helpful to know:
   (a) Are there any significant stresses occurring at home?
   (b) How is Brian doing in his math class?
   (c) Does Brian have stomach pain on the weekends?
   (d) Does Brian worry a lot?
   (e) Has Brian had his appendix removed?
3. The most appropriate initial therapeutic intervention for Brian might be:
   (a) Family therapy
   (b) Referral to the school counselor
   (c) Cognitive behavioral therapy
   (d) Psychiatric medication
   (e) Posttraumatic stress disorder treatment

Answers

1. (e): Symptoms that occur during school days and not on weekends and holidays

are not likely to be related to celiac disease and most likely to be functional in origin. Normal blood sugar results at the time of symptoms make a blood sugar related etiology unlikely.

2. (e): All of the other choices will help find the trigger for Brian's anxiety.
3. (b): Having the school counselor help Brian deal with his fears is the most appropriate initial intervention. Cognitive Behavioral Therapy can be an effective second line intervention, if necessary.

Case 2: Diabetes and Recent Weight Loss

Dan, a 15-year-old, with an 11-year history of diabetes, comes for a routine diabetes follow-up visit. Since his last visit 6 months previously, Dan's Hemoglobin A1C has increased significantly, reflecting poor blood sugar control. In addition, he has lost 12 pounds and appears undernourished. The blood sugars results recorded in Dan's log are lower than, and inconsistent with, his Hemoglobin A1C result. When given his test result, Dan's father states that he is disappointed because he "trusted" his son to be honest.

Dan is referred for evaluation and counseling. He admits to the social worker that he is feeling depressed and sometimes feels like hurting himself. He also feels that his diabetes has made him a "burden and disappointment" to his family. Dan is also upset that his parents have "backed off" and help him less with his diabetes regimen. Although they have told him that they are trying to be less involved, because he is now a young adult, he feels that they no longer want to deal with his diabetes.

Questions

1. The patient's weight loss can be explained by all of the following EXCEPT:
   (a) Elevated blood sugars
   (b) Depression
   (c) Decrease in appetite
   (d) Production of ketones
   (e) Low blood sugars
2. What is the best management for Dan at this point?
   (a) Referral for individual talk therapy
   (b) Referral for family therapy
   (c) Referral for immediate crisis intervention
   (d) Antidepressant medication
   (e) None of the above
3. What might Dan's parents have done to better transfer responsibility for diabetes management to Dan?
   (a) Requested that his physician order an insulin pump, so that he doesn't have to give himself injections
   (b) Had Dan renew and refill all his own prescriptions at age 12 years
   (c) Discussed Dan's need for independence and developed a mutually agreeable plan for increasing his diabetes-related responsibility
   (d) Had him come to his medical visits by himself
   (e) Gave him more responsibility for his diabetes but insisted that he text blood sugar results to his parents, each time he tests

Answers

1. (e): All can cause weight loss except for low blood sugars.
2. (c): Although medication, individual talk therapy, and family therapy are all effective treatments for adolescents with depression, suicide risk must be evaluated first.
3. (c): Developing a mutually agreeable plan for increasing Dan's diabetes responsibilities allows a gradual transition, keeping Dan safe as he develops more skills to care for his diabetes

Case 3: Diabetes and Difficulty Trouble Swallowing

Ann, an 11-year-old with a 16-month history of diabetes, is accompanied by her mother to an "emergency visit" with her diabetes nurse practitioner. Ann was reported to have had a "choking" incident two nights earlier and has been having trouble eating since, complaining that she feels

something in her throat that interferes with her swallowing. Ann feels that she choked as a result of her difficulty swallowing and has been refusing to eat anything solid. There is a strong family history of psychiatric illness. Ann's father takes medication for bipolar disorder and her mother and maternal aunt take medication for severe anxiety.

Ann is referred to a pediatric psychiatrist and is placed on antianxiety medication. The psychiatrist also refers Ann for cognitive behavioral therapy including exposure therapy (Chap. 19). Over the next month Ann responds to these interventions and begins to eat more, including solid foods.

Although improving, Ann still appears to struggle with her eating behaviors. Ann and her social worker continue to explore Ann's feelings, especially those related to her diabetes. Ann reveals that she has a fear of taking too much insulin and going "low." For this reason, she doesn't want to commit to what she is going to eat and administer her insulin dose before eating. This leaves Ann in a bind, with her choices limited to eating without receiving insulin and having high blood sugars, taking insulin after eating and risking a low sugar reaction, or skipping meals and snacks.

Questions

1. The following could all have caused Ann to have difficulty swallowing EXCEPT:
   (a) Elevated blood sugar
   (b) Posttraumatic stress disorder response
   (c) Psychosomatic reaction
   (d) Esophageal obstruction
   (e) Panic attack
2. Which of the following was the most influential factor used to determine Ann's initial treatment plan?
   (a) The strong family history of anxiety disorder
   (b) The patient had a possible eating disorder
   (c) The patient had a fear of low blood sugar
   (d) All of the above
   (e) None of the above
3. The best management for Ann's continuing disordered eating would be:
   (a) Increasing her antianxiety medication doses
   (b) Addressing Ann's fear of insulin overdose and low blood sugar reaction
   (c) Allowing her symptoms to improve spontaneously
   (d) Adding an antidepressant medication
   (e) All of the above

Answers

1. (a): Not being able to swallow is not associated with elevated blood sugar.
2. (a): A psychiatric referral was driven by the strong family history of anxiety.
3. (b): The underlying issues of the fear of the potential insulin over dosage and risk for low blood sugar need to be addressed before other options including psychotropic medication adjustment are considered.

Conclusions

Once medical causes of abnormal blood sugar levels, adherence issues, or atypical symptoms are ruled out, the clinician must consider the role of psychosocial factors. If a clinician only focuses on treating symptoms and does not consider their potential underlying psychosocial causes, they are less likely to improve and/or the diabetes is more likely to remain uncontrolled. Medical "fixes" such as increasing patient contacts, placing unrealistic expectations on patients and their families, and changing insulin doses when this not where the true problem lies are unnecessary and often frustrating to both families and caregivers. A more effective solution widens the focus to an approach incorporating collaboration between the diabetes care team, mental health professionals, and others in the community including teachers and school nurses. This systemic collaborative approach can improve blood sugar control as well as the overall wellbeing of the child or adolescent with diabetes and his or her family.

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Functional Somatic Symptoms in Pediatric Hematology and Oncology

10

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Abstract

Functional somatic symptoms (FSS) in pediatric hematology and oncology significantly limit health-related quality of life and functional abilities, thus requiring regular assessment and treatment. This chapter focuses on FSS in pediatric sickle cell disease and pediatric cancer, diseases for which there is an established literature describing and explaining functional abilities, health-related quality of life, and other adaptive outcomes. Pain and fatigue in pediatric sickle cell disease and pain, nausea, vomiting, and fatigue in pediatric cancer are among the most common and distressing disease and treatment complications. Although physical in etiology, psychosocial risks and resources across multiple levels of pediatric patients' social ecology influence the frequency, intensity, and course of these FSS. The literature regarding common approaches to assessment of pain, anticipatory nausea and vomiting, and fatigue as well as associated risk factors is summarized. Use of self- and parent proxy report measures of FSS, internalizing and externalizing symptoms, and psychosocial risk screening are recommended. Case studies demonstrate differential diagnosis and application of medical- and evidence-based psychological interventions for FSS. Importantly, multidisciplinary care models for pediatric sickle cell disease and pediatric cancer are consistent with the recommended approach to FSS.

Keywords

Pediatric cancer • Pediatric sickle cell disease • Pain • Nausea • Fatigue

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Functional somatic symptoms (FSS) have important implications for functional abilities and physical and emotional health-related quality of life in pediatric hematology and oncology [1, 2]. However, the high incidence of FSS in children and adolescents regardless of health condition [3], complicates assessment and intervention for FSS in children with chronic health conditions. For children with cancer or sickle cell disease (SCD), whose most common symptoms associated with disease and treatment are pain, fatigue, and nausea; assessment of FSS and determination of underlying physical and psychosocial factors are complicated. Health status varies widely across pediatric hematology and oncology diagnoses, but both cancer and SCD in childhood are marked by acute and chronic disease symptoms that significantly affect health-related quality of life (HRQL) or the subjective evaluation of physical, emotional, spiritual, and social functioning [4]. This chapter focuses on children with cancer or with SCD for whom there is an established literature setting the foundation describing functional abilities, HRQL, and adaptation and identifying factors that influence these outcomes.

Sickle Cell Disease

SCD occurs most often in individuals of African and Mediterranean ancestry [5]. There are several types of SCD; severity and symptoms vary by genotype, with homozygous sickle cell anemia (HBSS) and HBS beta thalassemia having the most severe pain and highest disease complications [5]. SCD is estimated to affect 70,000 Americans, including approximately 1 in 400 African Americans and 1 in 1,200 Hispanic Americans [6]. In the UK, the incidence is lower with approximately 140–175 infants born with SCD annually [7]. Sickle cell trait, a condition in which only one sickle cell allele is inherited, has been found in 2.0 % of newborns in Brazil [8] and 4.2 % of individuals prescreened prior to marriage in Saudi Arabia [9]. The rigid, sickle-shaped red blood cells in SCD can partially or completely occlude blood flow and in turn oxygen, to any part of the body, resulting in tissue and organ damage [6]. These unpredictable, vaso-occlusive events cause severe pain for the patient, varying in length from hours to weeks of chronic pain [10, 11]. Further, chronic organ damage involving the brain (overt or silent strokes), lungs, kidneys, and other organs can result in dysfunction and disability. SCD-related anemia, due to damage to red blood cells, can also result in fatigue, jaundice, and shortness of breath [6]. The multiple and chronic complications and unpredictable course associated with SCD can obscure understanding of FSS in SCD [11].

Psychosocial problems can occur for children with SCD due to several factors beyond the disease process such as disease-related shortened lifespan, stressors related to medical care of having family members with SCD, lack of access to specialized SCD care, and ethnic minority status and associated community stressors [12]. Delayed pubertal development as a result of SCD, chronic fatigue, and learning disabilities related to the neurocognitive sequelae of SCD may also serve to alter peer relationships of children with SCD [13]. Of note, classmates rate children with SCD as being sick more often and appearing more fatigued than their peers [14]. Because of the high incidence of low socioeconomic status among youth with SCD, at times, discrimination [15] and lower family education [12] can affect access to health care and the quality of care a child receives in the medical system. Previous research is mixed regarding the rates of externalizing symptoms (attention problems, conduct problems), internalizing symptoms (anxiety, depression, social isolation), and social competence (adaptive skills or resilience), though findings of limited HRQL of children with SCD are consistent (for a review, see Barakat et al. [12]). Because there are many risk factors for developing psychosocial problems, even in the absence of a diagnosis of a specific mental health disorder, there may be an increased rate of sub-threshold anxiety and depression within this population. On the other hand, children with SCD, who have not experienced a stroke have been found to exhibit "psychological hardiness" indicated by similar emotional well-being (assessed by symptoms of depression and loneliness,

positive self-concept), less aggressive behavior, and more prosocial behavior compared to healthy peers based on self-report and parent proxy report and teacher descriptions [13, 14].

Risk and resilience models have been employed to explain adaptive outcomes among children with SCD [16–20]. These risk and resilience models posit that coping, social and family resources buffer the impact of the disease and its complications on adaptation, while sociodemographic factors such as race/ethnicity and socioeconomic status can exacerbate stress associated with SCD. For example, in a series of studies, Barakat and colleagues found that maladaptive coping and high parenting stress are stronger contributors to reduced HRQL and increased disease complications than pain for adolescents with SCD [12, 21, 22]. Moreover, a bidirectional relationship between mood and pain has been identified, with higher anxiety and depression resulting in increased pain and increased pain resulting in higher anxiety and depression [23]. Further, low income has been associated with higher pain intensity and more functional disability among children with SCD [24].

Childhood Cancer

Cancer incidence for children under 15 is approximately 140 cases per million [25]. In the USA, cancer occurs in 1–2 of every 10,000 children [26, 27]. Although cancer is the leading cause of disease-related death in children, advances in childhood cancer treatments have dramatically reduced morbidity and mortality related to cancer over the last four decades. Five-year survival rates across all diagnoses are approximately 80 % [28], although survival rates vary dramatically by diagnosis. Treatment courses vary in length between diagnoses and typically include some combination of chemotherapy, corticosteroids (for chemotherapy or to treat cancer related symptoms), radiation, and surgery. Cancer and its treatments can lead to many side effects including nausea, vomiting, significant weight changes, fatigue, sleep disruption, headache, and neurocognitive changes during active treatment [11]. Importantly, there is increasing recognition of multiple chronic physical, cognitive, and psychosocial late effects of treatment that complicate the well-being of survivors of childhood cancer [29].

Acute lymphoblastic leukemia (ALL) is the most common childhood cancer, accounting for approximately 25 % of diagnoses under the age of 20 with a peak incidence between 2 and 5 years of age [27]. Advances in treatment have resulted in 5-year survival rates approaching 90 % for children with ALL [30]. Corticosteroids are a mainstay of chemotherapy for ALL and are known to disrupt sleep, increase fatigue, and affect mood and behavior [31]. Consequences of corticosteroids are also relevant to other malignancies such as brain tumors [32]. Brain and central nervous system tumors are the second most common childhood cancer, making up 16 % of pediatric cancer diagnoses, but unlike ALL prognoses are poorer and for those that survive, and treatment-related morbidity can be substantial [27]. Another group at great risk for poor health outcomes includes adolescent and young adult patients. Approximately 21,400 adolescents and young adults aged 15–29 years were diagnosed with cancer in 2000 in the USA, which is nearly three times the number of patients diagnosed in the first 15 years of life [33]. Internationally, prevalence rates are approximately 105–254 per million in adolescent males and 85–228 per million in adolescent females with considerable variation by country [34]. Symptom morbidity and mortality is also significantly higher for this age group relative to younger children [27].

Due to the sudden life threatening nature of a cancer diagnosis, acute stress reactions are often prominent around the time of diagnosis. Parent distress related to the cancer diagnosis is closely related to the child's distress during treatment and a positive family environment can reduce the impact of parental distress on child distress [35]. Cancer can involve multiple potentially traumatic events that together with the family's history and the subjective interpretation of the events can result in traumatic stress reactions. These reactions are defined as psychological and physiological stress responses such as heightened arousal to

stressful stimuli, re-experiencing traumatic events such as the diagnosis or specific procedures, and avoidance of discussion of topics, places, or people which are reminders of the cancer diagnosis; all of which can interfere with daily functioning [36]. Not all research, however, has confirmed the existence of posttraumatic stress in parents or children on active treatment [37]. Relative to peers without cancer, most research shows similar rates of anxiety and depression among children on treatment for cancer (see Patenaude [38] for a review) and cancer survivors [39]. Despite positive adjustment in many children and adolescents with cancer, approximately 25–30 % of patients experience clinically significant distress (see Patenaude [38] for a review), and for some, this is associated with functional symptoms. Risk factors for poor long-term adaptation include lower socioeconomic status, worse parental coping, and poor school performance [40].

The social ecological model has helped summarize factors that influence adaptation and HRQL among children with cancer [41]. This model posits that there are multiple influences on child development and adaptation that change over time from those most proximal to the child with immediate and continual influence, indirect influences, and those most distal from the child. Factors proximal to the child with cancer include characteristics of the child (e.g., age, presence of preexisting behavioral problems, or developmental disabilities), cancer (e.g., treatment intensity), and family (e.g., parenting stress, family functioning, prior family history of illness). Age at diagnosis can also impact psychosocial outcomes, as adolescents and young adults often face greater cancer morbidity and mortality than younger patients [42], while also experiencing many limitations on goal pursuit that is typical of same-age peers (i.e., getting a driver's license, applying to college) due to health status and at times a greater dependence on caregivers [43]. More distal factors include social and community supports and cultural influences. Family functioning variables, in particular, have been associated with HRQL [44]. Despite the many stressors faced during treatment, most children and families adapt well [45], and the literature describing positive outcomes from the cancer experience [46] including posttraumatic growth [47] continues to develop.

How to Make the Diagnosis of Functional Somatic Symptoms in Pediatric Hematology and Oncology?

The predominant approach to FSS in pediatrics incorporates prevailing models for explaining adaptation and psychopathology for children and adolescents with and without chronic health conditions [3]. The risk and resilience and social ecology models, which have been used to explain adaptation among children with cancer or SCD, can account for different symptoms, outcomes, and expression of FSS over time. These models summarize factors that influence FSS such as child age (more symptoms as children move into puberty), adaptive coping (coping strategies that bring about positive results such as problem-focused, positive thinking), family resources, and neighborhood quality/socioeconomic status. For example, Villalonga-Olives and colleagues [48] examined the association of life events to psychosomatic complaints (measured as headache, stomachache, backache, feeling nervous, sleeping difficulties, and dizziness). Results were supportive of the theory that somatization results from the experience of stress (a trigger) in the context of low socioeconomic, family, and social resources among adolescents and young adults. Specifically, family functioning mediated the association of negative life events with somatic complaints, such that more negative life events were associated with poorer family functioning which in turn predicted more somatic symptoms. Further, higher socioeconomic status buffered the effects of negative life effects on somatic symptoms (i.e., children from higher socioeconomic status families had fewer somatic symptoms regardless of presence of negative life events). Importantly, reciprocal relationships between internalizing symptoms (anxiety, depression) and FSS have been identified in both cross-sectional studies and with prospective data, suggesting that

anxiety and depression result in more FSS and FSS contributes to anxiety and depression [3]. Thus, children with internalizing symptoms are at risk for FSS and alternatively FSS put children at risk for poor adaptation.

In the case of pediatric cancer and SCD, symptoms are associated with disease processes and treatments [1]. Studies have sought to describe frequency, severity, and distress associated with a host of signs and symptoms in childhood cancer including hair loss, mucositis, eating problems, vomiting, and nausea. Pain, fatigue (a low energy state that persists despite adequate rest), and nausea (and other eating problems) are the most frequent and intense symptoms reported across studies of children and adolescents on active treatment for cancer [1]. However, in the case of cancer, few studies are comprehensive in their assessment of FSS among homogeneous samples of children (such as only children on treatment), and little is known about multiple symptoms and how they are associated with demographic characteristics and medical factors [1]. Pain is acknowledged as the most prevalent and disabling complication of SCD although chronic anemia with associated fatigue is endemic [49, 50].

Commonalities across pediatric cancer and SCD include the importance for explaining FSS of the child's social ecology or risks and resources of the child, family, school, and community (see earlier discussion of social ecological model) such as child age and development, child and family coping, family functioning, and sociodemographics. For example, parent involvement in disease management is greater for young children [51] suggesting that the role of parents in maintaining FSS is likely greater for young children. Adolescence and young adulthood present increased challenges for patients with SCD or cancer as this developmental period is characterized by rapid physical, cognitive, and psychosocial changes. Teens with SCD, who experience increased pain and functional disability [52], and teens with cancer, who face unique challenges in meeting developmentally appropriate goals [53], may experience increased health and psychosocial risks resulting in greater FSS.

Because FSS in pediatric hematology and oncology have a physiological basis as well as more subjective components, multiple measures completed by self and proxy reporters can be used to assess FSS. For both SCD and cancer, measures of health-related quality of life, in particular the PedsQL [54] and functional abilities such as the Functional Disability Inventory [55] are the first line in assessing whether FSS are influencing overall functioning. The Psychosocial Assessment Tool (PAT) is a brief screener of family risks and resources across categories of family structure, financial resources, patient and sibling behavioral problems, family functioning, and acute stress [40]. The PAT has been shown to be useful and feasible in both pediatric cancer [56] and pediatric SCD [57]. Use of symptom assessment scales in pediatric cancer have also been described [1]. Pediatric pain questionnaires are helpful in determining frequency, intensity, and location of pain [58] in both SCD and cancer, and valid and reliable measures that evaluate specific symptoms such as fatigue have been developed [59–61].

Case Studies

Case 1: Pain

John is a 19-year-old African-American male with homozygous sickle cell anemia who has a long history of disease complications including severe pain episodes resulting in extended hospital stays. He lives with his grandmother and three younger siblings. In addition to taking a course at the local community college, he works at a shipping company to support his siblings, but even with his income, the family continues to experience significant financial hardship. The stress John is under on a daily basis puts him at increased risk for pain [23] and can potentially impair preventative health behaviors such as eating a balanced diet, remaining hydrated, and having an opportunity for adequate sleep. Because of his frequent pain episodes, John needs increasingly larger doses of opioids for pain control. John has prescriptions for acetaminophen with

codeine and hydrocodone to help him manage the pain at home, but his pain often becomes so intense that only IV opioids will control his pain.

Recently, after 3 days of progressively worsening pain at home, John went to the emergency department alone. He explained to the triage nurse that he needed IV morphine because he had already tried 10 mg of hydrocodone every 3 h at home but the pain in his lower back was intense (a 9 on a 1–10 scale). The nurse rolled his eyes at John telling him that he is "clearly faking" and that he can wait for his "drugs" in the waiting room. John overheard the nurse telling other emergency department staff members that he was "drug seeking." Hearing the nurse call him a drug addict, John became very angry, but given his high levels of pain he could do little but wait. His anger further exacerbated his pain and made it difficult for John to employ adaptive pain-coping strategies. During a conversation with the pediatric hematologist on call, John reported that his pain remained at a 9/10 and that it was spreading from his lower back down to his legs making it extremely difficult for him to walk. The hematologist assessed John's pain history and how he has cared for it. The doctor admitted John to the hospital's hematology service and ordered morphine administered through patient controlled analgesia. John felt significant relief at the doctor's treatment plan and was then able to engage in relaxing and distracting activities that brought him increased comfort and reduced pain intensity in the past such as having family members visit, listening to music while engaging in deep breathing and imagery, and watching a favorite movie.

Questions

1. What steps can patients with sickle cell disease take to improve their emergency department visits?
   (a) Discuss a plan with their primary hematologist, including calling the hematologist on call before going to the emergency department.
   (b) Bring a family member to challenge hospital staff.
   (c) Bring documentation from their primary hematologist to the hospital stating diagnosis, current medications, and recommended treatment guidelines.
   (d) (a) and (c)
2. What pain management techniques can be used in the emergency department waiting room?
   (a) Heat packs and deep breathing.
   (b) Distraction, deep breathing, heat packs, hydration, oral medication, support from friends and family members.
   (c) Medication only.
   (d) Distraction with the television in the waiting room while trying to ignore the chaos of the emergency department.

Answers

1 (d) and 2(b): Because of the unpredictable nature of vaso-occlusive episodes that define SCD, some treatment is preventative, but the majority of treatment for SCD is palliative (i.e., treating symptoms and managing pain) in nature [62]. Approximately 90 % of pain episodes are managed in the home, in part, to improve the HRQL of those with SCD [63]. Cognitive-behavioral strategies such as deep breathing, progressive muscle relaxation, guided imagery, and calming self-talk are promising interventions to address sickle cell-associated pain in combination with pharmacologic intervention [64, 65]. Pharmacologically, SCD-related pain is typically managed with opioids, and patients can take increasingly larger doses of these medications to manage chronic pain [62]. Unfortunately, frequent opioid use can result in "concern-raising behaviors" that may be seen as drug-seeking behaviors (such as requesting specific medications or having high tolerance of opioids) by healthcare providers. Of note, however, no research has demonstrated evidence of substance abuse for those with SCD [15]. Cultural factors have also been shown to influence opioid prescription practices [66] and can be an important bias to address when working with patients presenting with pain. Furthermore, distraction, sleep, and interacting

with friends and family members are some of the most common non-pharmacological interventions reported by children, adolescents, and families [10]. Stress is closely related to the experience of pain in SCD and higher stress and more negative mood has been linked to increased pain ratings in children and adolescents [23].

Case 2: Anticipatory Nausea and Vomiting

Jake, a 12-year-old Caucasian 7th grader, was diagnosed with an optic pathway glioma after a period of headaches and visual changes noted during sports activities. Chemotherapy treatment was initiated but challenges ensued when Jake experienced significant nausea and vomiting secondary to chemotherapy. Although nausea and vomiting became controlled with use of antiemetic medication, by the third course of chemotherapy, Jake began to experience anticipatory nausea and vomiting in the car during the trip to the cancer center, entering the cancer center outpatient clinic from the elevator, and while being accessed for chemotherapy in the outpatient clinic. Referral to a psychologist occurred when Jake missed a scheduled chemotherapy and a follow-up visit in an attempt to avoid nausea and vomiting.

Assessment of psychosocial risk [67] and behavioral problems using a standard checklist indicated that Jake experienced mild anxiety and his family had a number of psychosocial risks for FSS. Contributing to risk for FSS for Jake, his recently divorced parents alternated bringing Jake to clinic, did not communicate directly with each other, and managed nausea in different ways. Jake's mother reinforced symptoms by allowing him to avoid clinic and miss treatments and his father set the expectation that Jake just needed to be "strong." Psychological intervention incorporated systematic desensitization with guided exposure (construction of an anxiety hierarchy based on geographical proximity to the cancer center outpatient floor from getting in the car at his house at the bottom of the hierarchy (least anxiety producing) to sitting in the day hospital receiving chemotherapy at the top of the hierarchy (most anxiety producing aspect of treatment), learning deep breathing and relaxation techniques, and use of positive scents (in this case a fabric softener sheet) during the trip to the hospital, on the elevator, and while on the floor). For scheduled chemotherapy treatment visits, oral antiemetics were started at home. Jake's anticipatory nausea and vomiting became more manageable and thereafter he was able to access his cancer treatment successfully.

Questions

1. Risks associated with FSS in pediatric hematology and oncology include:
   (a) Developmental considerations
   (b) Negative thinking and avoidant coping
   (c) Pre-existing family problems
   (d) All of the above
2. The most effective approach(es) to *anticipatory* nausea and vomiting is:
   (a) Prevention through early and effective use of antiemetics
   (b) Behavioral interventions to prevent symptoms development and/or to target symptoms once they have developed
   (c) None of the above
   (d) (a) and (b)

Answers

1 (d) and 2(d): Anticipatory nausea is common among children on chemotherapy treatment for cancer, occurring in up to 30 % of children [68]. Because anticipatory nausea and vomiting emerge through a process of classical conditioning or the association of chemotherapy induced nausea and vomiting with sights, sounds, and smells and experiences before, during, and after treatments, prevention through proactive use of effective antiemetics and hypnosis to "inoculate" against anxiety during procedures are first line approaches [69]. Behavioral interventions such as deep breathing, relaxation, hypnosis ([70], Chap. 21), and systematic desensitization (Chap. 19) have proven effective in reducing anticipatory nausea and vomiting when it develops [69, 71].

Case 3: Fatigue

Lilia is an 8-year-old, Asian-American girl diagnosed with intermediate risk pre-B cell ALL, now in the 4th month of maintenance treatment (the final phase of ALL treatment involving both oral and intravenous chemotherapy). Lilia is the youngest daughter of three children of a widowed mother. The maternal grandmother lives with the family and cares for the children while mother is at work; she is highly anxious about Lilia's recovery and ultimate prognosis. Lilia experiences irritability and fatigue that extends for 1–2 weeks following each 5-day prednisone treatment. Lilia demonstrates heightened sensitivity to steroids [72] and her significant behavioral symptoms are pronounced throughout the washout period as the steroids leave her system [73]. As a result, Lilia spents much of her day watching television or on the computer, was unable to return to school, and was not engaging with peers or siblings. Sleep also was disrupted, and her mother often stayed with her most of the night to help her fall and return to sleep.

Multidisciplinary treatment [74] included parent education and support (with mother and maternal grandmother) to create a schedule for schoolwork, chores, and activities with associated reinforcement for effort. In addition, sleep intervention incorporated maintenance of a daily sleep diary [75] by her mother with introduction of healthy sleep hygiene (including regular bedtime/wake time, sleeping in a sleep conducive environment, use of a bedtime routine, and reducing daytime sleep) [76, 77] to facilitate quality sleep. Graduated extinction (incrementally reducing Lilia's mother's presence and involvement at bedtime slowly over a period of several days to weeks) to promote Lilia's ability to sleep independently was implemented [76, 77]. Increased daily physical activity to improve physical strength by having Lilia and her grandmother take a walk around the block each morning was prescribed as part of treatment. Within 6 months, ratings of fatigue and of health-related quality of life were within the normal range. Lilia had returned to school with appropriate accommodations to support her during steroid pulses, was working consistently on her schoolwork and achieving better grades, and had joined the local scouting group.

Questions

1. Fatigue in pediatric cancer is most often associated with:
   (a) Pain
   (b) Pain medication
   (c) Chemotherapy treatment including corticosteroids
   (d) Folic acid deficiency
2. Resilience in pediatric SCD and pediatric cancer is associated with:
   (a) Low parenting stress
   (b) Child behavior problems
   (c) Social supports
   (d) (a) and (c)

Answers

1 (c) and 2(d): Increased fatigue is related to both corticosteroids [78, 79] and chemotherapy treatments, typically decreasing as the child gets further away from the chemotherapy treatment [80]. Corticosteroid-related sleep disruption can further exacerbate fatigue [78] making it difficult for children to remain active and keep consistent sleep wake schedules (i.e., not nap during the day). Behavioral sleep interventions focus on maintaining good sleep hygiene (e.g., regular bed and wake times, no additional naps, using bedtime routines) while also extinguishing problematic sleep onset associations (learned behaviors a child needs to fall asleep—for example needing a parent present to fall asleep) during steroid bursts [76, 77]. Helping families maintain daily schedules, family routines, and consistent expectations of their child during treatment is essential to managing steroid-related behavior disruptions as well as regular sleep habits.

Treatment Summary and Conclusions

Pain and fatigue in pediatric SCD and pain, nausea, vomiting, and fatigue in pediatric cancer are among the most common and most distressing disease and treatment complications. Although physical in etiology, psychosocial risks and resources across multiple levels of pediatric patients' social environment influence the frequency, intensity, and course of these FSS. Importantly, FSS in pediatric hematology and oncology significantly limit quality of life and functional abilities and require regular assessment and treatment. Although this chapter does not describe symptoms of fatigue and pain at end of life, there is adequate evidence to suggest that more is needed to treat and reduce suffering in this period [81].

Central to comprehensive cancer and SCD care is the provision of integrated psychosocial interventions. Multidisciplinary care is also key to addressing FSS and important for differential diagnosis and determination of treatments. Sharpe and Carson [82] are proponents of a "paradigm shift" in which psychological treatments are integrated into medical care in an effort to more directly evaluate and address FSS. An example of multidisciplinary care for childhood cancer is the psychosocial services program of a number of children's cancer centers in which psychosocial staff such as social workers, child life specialists, teachers, spiritual counselors, creative arts therapists, psychologist and psychiatrists, partner with physicians and nurses to provide support, cancer education, school reentry programming, and intermittent minimal financial support to all children and their families as well as specialized care for those with specific risks such as intervention for children with anxiety, neuropsychological assessment for children with brain tumors, and age appropriate activities and support for adolescent and young adult patients [83]. Comprehensive sickle cell centers have also implemented this multidisciplinary approach to care involving health educators, social workers, and psychologists in the services provided to their patients and families [84].

In conjunction with a medical exam, use of self- and proxy-report measures of FSS as noted in "How to Make a Diagnosis of FSS" and associated risks factors can aid in differential diagnosis by identifying the unique set of risks and resources presented by the patient and family. For example, understanding a family history of SCD or of cancer and prior experiences with the medical system for the patient and family members may be critical for understanding development and expression of FSS. Also, cultural background may influence experience of and communication about FSS [50].

For intervention, clinicians should consider (1) cognitive-behavioral treatments of underlying anxiety and depression (Chap. 19), especially for those with multiple FSS; (2) education to present information about appropriate disease management and the importance of adherence for effective management of complications and treatment of disease; and (3) parenting and family interventions to address parents' roles in maintaining symptoms and support parenting that promotes functional abilities and HRQL [85]. Addressing risks and resources (e.g., social support in the community and/or school system) in the broader social ecology such as financial barriers to care, family problems, and school issues that contribute to FSS should also be considered in developing a treatment plan. Thus, multidisciplinary care models, considered the standard of care for pediatric SCD [86] and pediatric cancer [41, 87] are consistent with the recommended approach to FSS.

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Chronic Pediatric Rheumatologic Pain Syndromes

11

Murray H. Passo and Lara H. Huber

Abstract

Chronic and recurrent musculoskeletal pain in children and adolescents is a common problem for primary care physicians to address. These pain syndromes are categorized in the spectrum of biopsychosocial syndromes that incorporate physical symptoms and signs with psychological components, often amplified by environmental or social influences through partially understood mechanisms. The patient may experience localized or widespread pain. The syndromes can be readily recognized by patterns of symptoms and signs that are illustrated in the chapter. Reflex sympathetic dystrophy, fibromyalgia syndrome, and benign hypermobility syndrome have physical findings that help to define the diagnosis; while growing pains are void of objective physical findings but are defined by the episodic nature and characteristic history. There is often an association with excessive school absences, anxiety, depression, and dysfunctional relationships with the parents. The characteristic patient profiles and the cardinal findings are illustrated. It is important to diagnose these pain syndromes early and institute therapy in order to avoid unnecessary investigations, identify contributing factors, improve function, and render pain relief. Most patients are amenable to treatment; however, recurrences are common, and underlying emotional contributions are difficult to eliminate.

Keywords

Reflex sympathetic dystrophy • Complex regional pain syndrome type 1 • Reflex neurovascular dystrophy • Fibromyalgia • Growing pains • Benign hypermobility syndrome • Pain centralization syndromes • Pain amplification syndromes • Chronic idiopathic musculoskeletal pain syndromes

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Introduction

Evaluation of musculoskeletal pain is a common problem for the primary care physician. A recent literature review of the epidemiology of chronic pain in children and adolescents demonstrated that chronic musculoskeletal pain in children and adolescents is very prevalent with a wide range, 3.9–40 % [1]. Chronic pain can be either recurrent or persistent and is typically defined as being present for 3 or 4 months. The wide range of prevalence rates likely is related to an assessment of patient population that includes different age groups and use of different definitions of chronic pain. It is more common in girls as compared to boys and increases in prevalence with age. A study of 749 healthy German children, elementary school ages 6–9 years old and secondary school ages 10–18 years old, reported nearly 11 % of participants had limb pain and 9 % had back pain [2]. There was significant associated disruption of sleep, school attendance, daily activities, appetite, and recreational activities [2].

The economic impact on society and the economy in the UK was estimated by Sleed et al. to be equivalent to billions of dollars of healthcare expenses [3]. Thus, the impact on the child, as well as the family, time off work, and out-of-pocket expenses is astounding. This is a serious health issue and requires improved diagnosis, expertise in management, and empowerment of the patients and parents for improved outcomes. Proper identification of the pain syndrome allows more focused treatment in the appropriate channels and avoids excessive testing and inappropriate medications.

Pain may be triggered by a defined illness or trauma but may evolve to become a chronic condition with no explained medical disease. The mechanisms of chronic pain mediation are complex and can be divided into peripheral and central mechanisms [4]. These mechanisms are beyond the scope of this chapter and for more information the reader is referred to the excellent reference by Lee [4]. Fibromyalgia is the prototype of a chronic widespread pain syndrome and is considered to be a result of central sensitization because of a disturbance in pain processing, which leads to pain amplification. Thus, this type of pain is mediated differently than acute pain. Yunus provides an excellent review of the central sensitization syndromes that incorporates fibromyalgia, irritable bowel syndrome, chronic tension and migraine headaches, restless legs syndrome, interstitial cystitis, temporomandibular joint syndrome, and chronic fatigue syndrome [5].

The idiopathic musculoskeletal pain syndromes, commonly referred to as pain amplification syndromes, can be divided into localized and diffuse [6]. Some patients have features of both patterns:

(A) The localized pain group refers to Reflex Sympathetic Dystrophy (RSD), now known as Complex Regional Pain Syndrome Type 1 (CRPS1). Historically it has been called Reflex Neurovascular Dystrophy (RND), post-traumatic pain syndrome, and in the adult, Sudeck atrophy.

(B) The diffuse pain group refers to fibromyalgia that has been recognized in adults for decades but has also been further described and diagnosed in children and adolescents, in whom it has been named pediatric fibromyalgia or primary juvenile fibromyalgia syndrome [7, 8].

Another group of children and adolescents have chronic idiopathic musculoskeletal pain that lacks the pathognomonic features of juvenile fibromyalgia or RSD and does not fulfill the diagnostic criteria for these diagnoses.

Two important pain syndromes that are common and need to be readily diagnosed are “growing pains” and benign hypermobility syndrome. These two pain syndromes have potential for pain amplification and require proper identification and patient education. When confronted with symptoms of musculoskeletal pain, the physician must determine which complaints suggest underlying malignancy or an inflammatory or autoimmune disease. The description of the pain allows pattern recognition, and the physical examination helps to distinguish serious diseases from idiopathic pain syndromes in most cases. Recognition of these pain syndromes will obviate excessive testing and facilitate targeting therapy in the appropriate areas.

Idiopathic musculoskeletal pain syndromes implies that the etiology is undetermined; moreover, the origin is likely multifactorial. There are several clues that suggest a predisposition to developing chronic pain. Malleson has described "intrinsic" factors that predispose to chronic pain syndromes including low pain thresholds, female gender, hypermobility, poorly perceived control over pain with maladaptive coping strategies, and "difficult" temperament. "Extrinsic" factors contributing to the chronic pain include previous pain experiences, social deprivation, physical or sexual abuse, parental modeling of chronic pain behaviors, sleep disturbance, and decreased physical fitness [6]. The common features of these patients are female gender, early adolescent, extremely high pain rating with a visual analog scale (VAS) 9-10/10 (commonly exaggerated, e.g. "100/10"), pain unresponsive to therapy, and marked functional disability such as missing numerous days of school or requiring homebound instruction. Some patients exhibit conversion symptoms [9]. It is also important to recognize that patients with well-defined diseases, such as juvenile idiopathic arthritis or systemic lupus erythematosus, can also develop secondary pain amplification syndromes. Diagnosis of the pain amplification will target the intervention at the underlying secondary pain rather than at a "presumed" flare of the primary disease, though this can be very challenging. This has significant ramifications in terms of treatment efficacy and avoidance of potential excessive medications with potential adverse effects. This concept has been recently reviewed in the context of adult rheumatic diseases and is germane to children as well [10].

Psychological contribution to the pain, either primary or secondary, also has been suggested by several investigators. It is important to evaluate for role models of chronic pain, psychosocial stressors, pre-existing psychiatric diagnoses, school performance, learning difficulties, significant life events, and presence of generalized pain [11]. Sherry et al. reported 100 children with psychosomatic musculoskeletal pain and found two predominate patterns of family milieu; one chaotic and one cohesive. The chaotic group experienced more distress. Interestingly, depression was found in only 11 % of the patients. Enmeshment between the parent (usually the mother) and the patient, defined as an inappropriate closeness and involvement of the child in the parental affairs and vice versa, was demonstrated in 76 %. Although many patients were characterized as "bright," on average they were found to have normal intelligence and some had school difficulties [12].

Case scenarios of patients with pain amplification syndromes are illustrated below to provide the cardinal signs and symptoms, thus allowing pattern recognition.

Benign Hypermobility Syndrome

Case 1

The patient is a 14-year-old Caucasian male who presents with joint pain since the age of three. He has pain in most of his joints including his ankles, knees, hips, wrists, elbows, shoulders, and hands. On his examination, he had hyperextension of the bilateral knees to 25°. In addition, hypermobility of the ankles, hips, wrists, elbows, and shoulders was noted. His skin was also found to be soft. He was diagnosed with benign joint hypermobility syndrome (BJHS) also known as Ehlers–Danlos Syndrome-hypermobility type (EDS-HM). He was treated with nonsteroidal anti-inflammatory drugs, referred to physical therapy, and encouraged to continue low-impact physical activity. Despite these interventions, the patient continued to have pain that progressed to chronic pain amplification. He subsequently developed anxiety associated with school, and he was placed on homebound status.

Joint hypermobility is a relatively common phenomenon among children. The prevalence varies and has been reported to be between 2.3 and 30 % [13–20] depending on the age and ethnicity of the population. Hypermobility is more commonly present in females, [13, 21, 22] and its presence decreases with increasing age [21, 23]. A distinction must be made between children with generalized joint hypermobility and BJHS. Generalized joint hypermobility is diagnosed

Table 11.1 The Beighton Score

Maneuver	Points (0–9) Right	Left
Passive dorsal flexion of the little finger of more than 90° with the wrist in the mid position[a]		
Passive movement of the thumb so that the thumb touches the ventral side of the lower arm[a]		
Active extension of the elbow of more than 10°[a]		
Active extension of the knee of more than 10° lying on the back[a]		
Bending forward with stretched knees so the palms touch the ground[b]		
Total		

[a]One point for each side
[b]One point

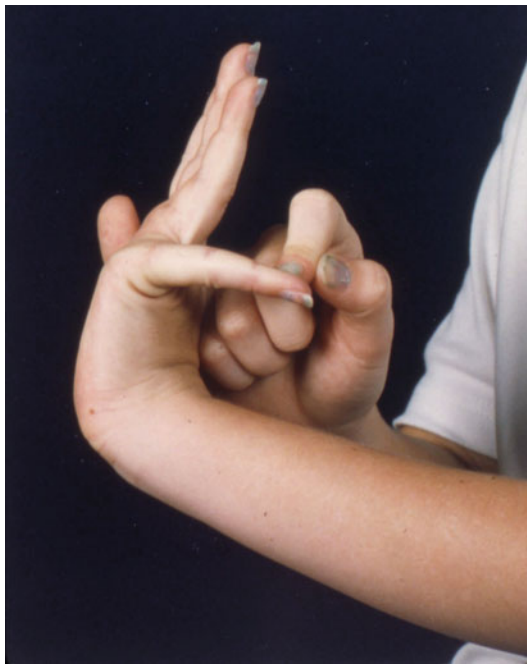

Fig. 11.1 Passive dorsal flexion of the fifth digit to >90°

based on physical exam using a Beighton Score. There is some controversy as to the score that should be used for diagnosis in children although a score of 5–7 or greater is generally accepted as abnormal [23–26] (Table 11.1, Figs. 11.1, 11.2, 11.3, 11.4, and 11.5). BJHS or EDS-HM is diagnosed based on criteria that were revised in 1998

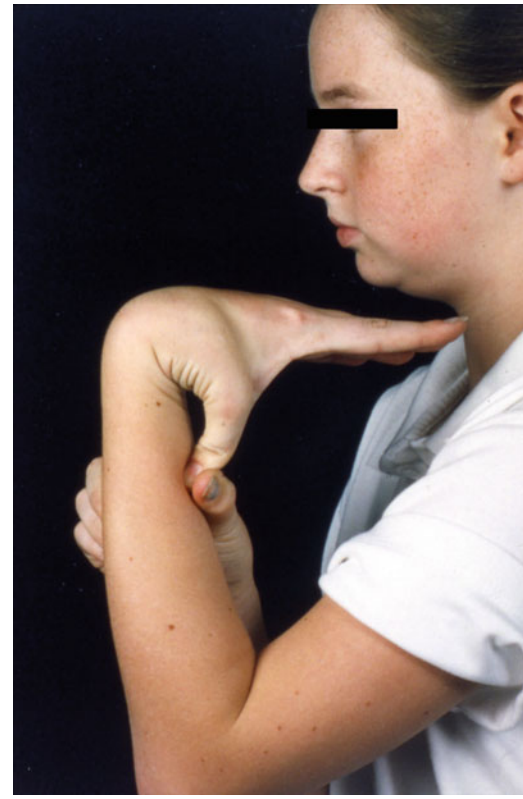

Fig. 11.2 Passive movement of the thumb so that the thumb touches the ventral surface of the arm

(The Brighton Criteria) [27] (Table 11.2). The Brighton criteria have not been validated in children, so the results should be interpreted with caution when used in children under 16 years of age [27].

EDS-HM type or BJHS is a clinical diagnosis and cannot be diagnosed with genetic testing unlike other types of Ehlers–Danlos. Children with BJHS will often have arthralgia, a history of joint dislocation, a Marfanoid habitus, and hyperextensible skin [28]. Contrary to its name, BJHS is not benign. It has been associated with a host of other ailments which, while not dangerous, are quite troublesome and disruptive, such as fibromyalgia, pain amplification, and centralization of pain [29–31]. In a recent study by Ting et al. nearly half of 131 children with juvenile fibromyalgia were found to have joint hypermobility [31]. A case–control study published in 2012 found that children with hypermobility syndrome were

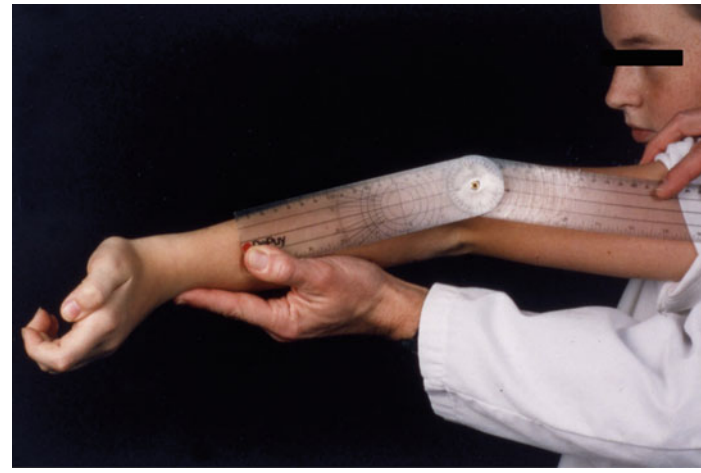

Fig. 11.3 Hyperextension of the elbow to ≥10°

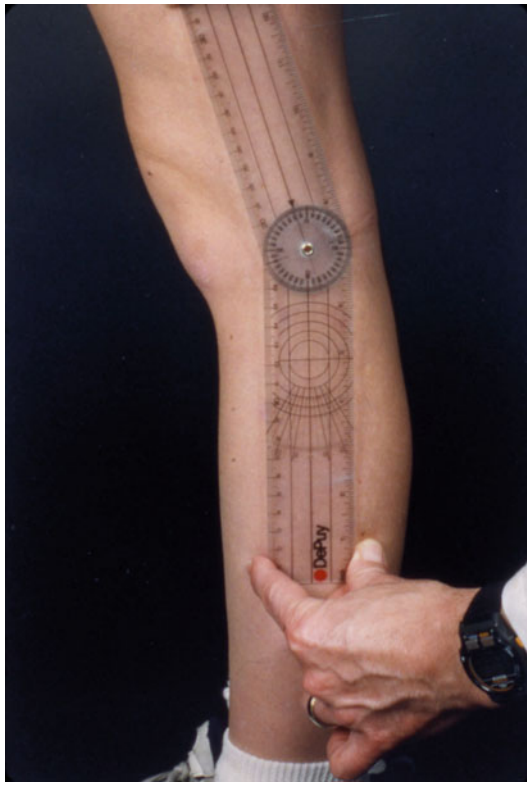

Fig. 11.4 Hyperextension of the knee to ≥10°

Fig. 11.5 Bending forward with stretched knees so that the palms touch the ground

found to have higher pain intensity and decreased quality of life [32]. There is a significant association between anxiety and joint hypermobility [33–35]. In a case–control study published in 1998, 67.7 % of patients with anxiety disorder were found to have hypermobility as compared to 10.1 % in psychiatric controls without a diagnosis of anxiety disorder and 12.5 % in medical controls [35]. Bendik et al. found that 75 % of 28 women with joint hypermobility syndrome suffered from migraines as compared with 43 % or healthy controls [36]. Autonomic symptoms such

Table 11.2 The Brighton (1998) criteria

Major criteria
1. A Beighton score of 4/9 or greater (either currently or historically)
2. Arthralgia for longer than 3 months in four or more joints
Minor criteria
1. A Beighton score of 1, 2, or 3/9 (0, 1, 2, or 3 if ≥50 years of age)
2. Arthralgia (≥3 months) in 1–3 joints, or back pain (≥3 months), spondylosis, spondylolysis/spondylolisthesis
3. Dislocation/subluxation in more than one joint, or in one joint on more than one occasion
4. Soft tissue rheumatism ≥ 3 lesions (for example: epicondylitis, tenosynovitis, bursitis)
5. Marfanoid habitus: tall, slim, span/height ratio >1.03, upper:lower segment ratio <0.89, arachnodactyly (+Steinberg/wrist signs)
6. Abnormal skin: striae, hyperextensibility, thin skin, papyraceous scarring
7. Eye signs: drooping eyelids or myopia or antimongoloid slant
8. Varicose veins or hernia or uterine/rectal prolapsed

Diagnosis = two major criteria, one major and two minor criteria, or four minor criteria
Two minor criteria are adequate if a first degree relative is affected
A diagnosis of Marfan syndrome or Ehlers Danlos Syndrome (other than the hypermobility type) excludes diagnosis of BJHS

as syncope, presyncope, orthostatic hypotension, postural orthostatic tachycardia syndrome (POTS), palpitations, chest discomfort, and heat intolerance have also been found to be more common in patients with joint hypermobility [37]. A gender matched case–control study found that children with chronic fatigue syndrome were more than three times more likely to have joint hypermobility than healthy controls [38].

Diagnosis of BJHS is made largely based on clinical presentation including both the history and physical exam. The most common presenting symptom is arthralgia, most often in the knees or ankles [13]. Other presenting symptoms include gait problems, lack of coordination, back pain, problems with handwriting, or clicking joints [13]. Most patients will complain of pain exacerbated by exercise, either immediately following the exercise or later in the evening or the following morning. Joint problems may include swelling, reduced range of motion in joints, stiffness, sprains, foot pain, growing pains, and, dislocations or subluxations [13, 25, 39]. While a reduction in joint range of motion may seem contradictory, decreased range of motion is caused by injury resulting in inflammation and effusion in a hypermobile joint, which in turn may lead to reduction in range of motion. The effusions in hypermobile joints are generally episodic and shorter in duration than those seen in inflammatory diseases such as juvenile idiopathic arthritis that are persistent until treated. Because of the associations mentioned above, children with BJHS often complain of fatigue, disordered sleep, syncope, palpitations, anxiety, and depressed mood. Parents frequently give a family history of a first-degree relative with hypermobility [13, 25]. Sixty-three percent of 125 patients with BJHS were found to have a first-degree relative with joint hypermobility in a study published in 2005 [13].

The physical exam is essential in identifying hypermobile joints. The physical exam should also be used to rule out other more serious conditions. Special attention should be given to the skin exam, heart exam, eye exam, and body habitus to identify signs that may indicate another condition such as Marfan syndrome or Ehlers–Danlos syndrome (aside from the hypermobility type). Laboratory examination is unnecessary unless another condition is suspected.

Treatment must begin with educating the patient and the patient's family regarding the condition. Emphasis should be placed on the episodic nature of the condition despite the presence of troubling symptoms [25]. Physical therapy is usually employed and is aimed at strengthening the muscles supporting hypermobile

joints, proprioception training, and improving stamina [25, 40]. A randomized control trial published in 2010 found no difference in patient pain scores between a targeted physical therapy program targeting muscles surrounding symptomatic joints and a general physical therapy program focusing on overall fitness. However, both groups showed an improvement in pain [41]. Occupational therapy may also be helpful in patients who complain of pain with writing or poor handwriting [25]. In patients who are also plagued with pain amplification or centralization of pain, then referral to a clinical psychologist for cognitive behavioral therapy is indicated (Chap. 19) [25]. Patients with anxiety or depression should be evaluated and treated with psychotherapy or pharmacologic agents (Chap. 26) [42]. Good sleep hygiene should be emphasized, and a pharmacologic sleep agent should be considered on a patient-specific basis.

Growing Pains

Case 2

The patient is a 5-year-old female who presents with episodes of leg pain since the age of 3 years. Her parents describe severe pain in both legs occurring mainly over the shins. The pain occurs two to three times per week, and it will often cause her to cry. The episodes may last up to 3 h. The pain is improved by massage and usually responds to analgesics such as acetaminophen or ibuprofen. It resolves by the following morning. Her parents have not noticed any swelling, warmth, or redness over the painful areas. She is growing well and has no other complaints. Her parents have noticed that the episodes usually occur after she has had a particularly active day, sometimes awakening her from sleep. The physical examination is entirely normal. She was diagnosed with growing pains. No further investigations were undertaken. Her parents were instructed to continue supportive care as they have been doing.

Growing pains are a common cause of pain in childhood. The reported prevalence of growing pains varies greatly ranging from 2.6 to 49.9 % [43, 44]. This variation is due to differences in the age and ethnicity of the populations studied in addition to inconsistency in the criteria used to define growing pains.

Although the term "growing pains" has often been loosely used to refer to idiopathic limb pains occurring in children, this term should be defined fairly specifically. Growing pains are usually non-articular pains that may be severe in nature. These occur deep in the bilateral upper or lower extremities without evidence of swelling, inflammation, injury, or an underlying systemic condition. Growing pains usually occur in the afternoon or at night and resolve by morning [45–47]. In a series of 30 patients, 43.3 % had pain at night and 56.7 % had pain in the afternoon [48]. The frequency of the pain varies from daily to every few months [48, 49], and the most common sites of pain are in the shins and thighs [48]. Episodes of crying with pain are frequently reported, and severe episodes often follow times of intense physical activity [48, 49]. Recent studies have used the above definition to evaluate the prevalence of growing pains. Evans et al. reported a prevalence of 36.9 % in a survey study of 1,445 children in South Australia in 2004 [50]. In 2009, a retrospective study of 531 Mediterranean children reported a history of growing pains in 24.5 % of the study population [49].

Making a diagnosis of growing pains may be difficult primarily because other more serious causes of limb pain must be considered and excluded. The physical exam should be conducted very carefully and include a thorough musculoskeletal exam including examination of all joints and muscle strength testing. It should focus on identifying abnormalities that might suggest an underlying organic cause for the pain. A diagnosis of growing pains should not be made unless the physical exam is normal. One needs to be extra careful in the patient with unilateral complaints since that situation heralds concern for an alternative diagnosis. Laboratory tests and imaging are not necessary to make the diagnosis; however, if an underlying condition is suspected, then laboratory examination and imaging may be warranted. A sex-and age-matched case–control study published in 2007 included 100 patients between the ages of 3 and 15 with recurrent,

intermittent limb pains lasting >3 months. Exclusion criteria included (1) any abnormal systemic symptoms and signs, (2) localizing signs including tenderness, swelling, warmth, or erythema, (3) joint involvement including swelling, pain, redness, or warmth, and (4) limp or limitation of activity. Laboratory evaluation included a CBC, ESR, calcium, phosphorus, alkaline phosphatase, and rheumatoid factor. No clinically significant difference was found in laboratory results between patients and 150 sex- and age-matched controls [51].

The etiology of growing pains remains unclear. An association between headaches and growing pains has been reported [52] raising the question of a decrease in vascular perfusion being an etiologic factor in growing pains. However, a significant difference in vascular perfusion patterns between children with and without growing pains was not found. In addition, there was no difference in vascular perfusion patterns in children affected by growing pains at times that they reported pain as compared to times when they did not report pain [53]. Hashkes et al. found that 44 children with growing pains had a lower pain threshold than healthy controls [54]. In a 5-year follow-up study of these 44 patients with growing pains, about half of the cases of growing pains had resolved. In the 17 patients with persistent growing pains, the majority had a decrease in the frequency and severity of their symptoms. Only three patients had worsening of their symptoms. The patients whose pain resolved were found to have improved pain thresholds, whereas patients with persistent pain continued to have low pain thresholds as compared to healthy controls. None of the patients developed juvenile fibromyalgia. The authors concluded that growing pains generally carried a benign prognosis and likely represent a pain amplification syndrome of childhood [55]. It has also been noted that growing pains are more prevalent in children whose parents experience pain [45, 56]. In a case–control study published in 1951, Naish and Apley observed that children with limb pains had more "minor nervous problems," such as irritability, nervousness, fear of the dark, bad dreams, nightmares, nocturnal enuresis, and tics, as compared to children without limb pains [56]. These observations further support the theory that growing pains may be a pain amplification syndrome of childhood [52, 56]. There also seems to be an association with decreased bone density in children with growing pains. In a study published in 2005, Friedland et al. found a reduced bone density in children with growing pains as compared to healthy controls [57]. A 5-year follow-up of these patients found a correlation between increase in bone density and improvement in growing pains [58]. Furthermore, an association of hypovitaminosis D with growing pains has been suggested [59].

Treatment is primarily supportive care and education to the parents and patients regarding the benign nature of growing pains. Many parents report that massage is a successful technique [49]. Over the counter analgesics, such as acetaminophen or ibuprofen, may be used [49, 60]. If the child suffers from severe, persistent growing pains, then a longer acting NSAID such as naproxen may be given at bed time on a scheduled basis [54, 60]. Analgesics may also be given as needed at bedtime if an episode of growing pains is anticipated because of a day with increased physical activity [60]. Despite the association of low levels of vitamin D [59] and lower bone density [57] with growing pains, no evidence to support calcium or vitamin D supplementation has been reported to date.

Reflex Sympathetic Dystrophy, Reflex Neurovascular Dystrophy, Complex Regional Pain Syndrome Type 1 (CRPS1)

Case 3

The patient is a 15-year-old female who was seen in the office with a swollen, exquisitely painful left foot. The patient was seen 3 weeks previously in a local Emergency Department and thought to have a Salter 1 fracture of the distal fibula secondary to trauma; however, the patient was unable to recall a specific traumatic incident. She was placed in a walking cast but the pain intensified and she requested the cast be removed.

She had taken ibuprofen and some oxycodone, left over from a tooth extraction, which did not help the pain. On physical examination she had a cool, pale, edematous left foot and ankle which was extremely sensitive to light touch. The patient refused to bear weight on the foot. She was felt to have complex regional pain syndrome and promptly referred for physical therapy. She fully recovered in a few weeks; however, she was subject to brief recurrences.

RSD is widely recognized in adult patients as a post-traumatic phenomenon; however, overt major trauma is not common in children with this syndrome. There may be a history of minor, insignificant trauma although this is often lacking. Psychosocial contribution is reported in many series of pediatric RSD/RND/CRPS1 (the latter term is now preferred). The pain is usually in the distal lower extremity; however, it occurs in the upper extremity as well and can rarely occur in other areas. The classic syndrome presentation is in a patient who has a swollen, diffusely puffy distal limb, intense burning pain at the affected site, and vasomotor changes from autonomic instability. Physical examination reveals the site is either warm or cold, red, or pale, with excessive sweating, marked hyperalgesia (increased sensitivity to noxious stimuli), and allodynia (pain provoked by stimuli not usually considered painful, such as light touch). The child holds the limb motionless as if paralyzed and resists any movement. Immobilization for suspected fracture or severe sprain actually aggravates the pain. Jacobs describes this presentation as hysterical edema, emphasizing the pseudoparalysis and subconscious effort not to move the body part [61]. In a series from the Netherlands, Tan et al. compared 78 children to 951 adults with CRPS I. Significant differences were the following: the children were mostly female, were more likely to have lower extremity involvement, had mostly colder temperature of the affected limb, and had less edema as compared to adults. In children with CRPS I, 62 % occurred after minor trauma. Long-term prognosis was good functional return, but recurrences were seen in 28 % of the children, particularly with new injury [62]. There was no reference to psychosocial contribution in that series.

Bernstein et al. published a series with 24 episodes in 23 children in 1978, referring to the diagnosis as RND [63]. Lower extremity involvement was present in 20/23 patients, 18/23 were girls, exquisite tenderness was seen in all affected limbs, swelling was present in 19/23, and vasomotor instability was present in 21/23. Coolness of the limb was found in 18 of those 21 and discoloration in 13 of those 21. Psychological findings included parental conflict, marked tendency to accept responsibility beyond years, and over involvement in school. No dystrophic changes were found. Physical therapy was employed as the treatment with good success, although minor relapses or recurrences of pain were reported in seven patients.

Sherry reported psychosocial factors in 21 children with RND [64]. He found parental/child enmeshment in all, significant school problems in 13, marital discord in 12, predominant high internal cohesion in 15 families with low levels of conflict, and 6 with low levels of cohesion but high levels of obvious conflict. Possible role models in the family for chronic pain were present in ten patients. This study emphasized the psychosocial contribution to the pain amplification syndrome, thus requiring therapy to address this stress related condition.

Stanton et al. reported 49 episodes of RSD in 36 children. In the 23 patients who had psychological testing, 83 % were found to have emotional dysfunction, including but not limited to divorce of parents and physical or sexual abuse [65].

In a report of 17 pediatric patients with CRPS 1 in an inpatient setting, the patients were studied for emotional and neuropsychological profiles. Projective personality testing found that 65 % supported anxiety or depression, 60 % had somatization, and 67 % had family and peer problems. Of 11 patients tested, 36 % were at risk/impaired on the attention/impaired working memory composite. The results showed generally intact cognitive functioning [66]. The authors support the concept of emotional distress in this group of patients.

First-line treatment of the condition requires physical therapy to desensitize and move the painful limb. Desensitization includes rubbing with textured fabric, contrast baths, and hand

massage. Many patients improve with physical therapy alone; however, psychological interventions are utilized in many patients as well. A recent systematic review of 12 studies reported that there was poor-to-fair quality evidence on the short-term benefit of physiotherapy for children with CRPS in combination with other modalities [67]. This detailed review of the evidence showed that the numbers of studied patients were generally small and the outcome measures were variable. More studies of high quality were recommended. Consultation with a pediatric rheumatologist is helpful to orchestrate the physical therapy and recommend medication if necessary. A fraction of the patients will need a pain medicine consultation for sympathetic block or more aggressive pain management.

In a large series from Boston, Wilder et al. presented 70 patients who were treated based on an algorithm that included a multidisciplinary team and multiple treatment modalities [68]. All patients received physical therapy. Some of the patients received transcutaneous nerve stimulation, cognitive behavioral therapy (Chap. 19), sympathetic nerve blockade, and medications. Psychological assessment was performed to diagnose underlying conditions that may inhibit treatment success. Forty four received therapy from behavioral medicine, and 57 % reported benefit from the psychological treatment. Conservative, noninvasive treatment was successful in 40, and 37 received the sympathetic blocks. Thirty-eight of the 70 patients had residual pain and loss of function.

Sherry reported successful outcomes in 92 % of 103 patients treated with intense physical and occupational therapy (PT/OT), defined as 4–6 h per day for a mean of 14 days, and psychological counseling. No patients required invasive treatments such as sympathetic blockade [69]. A prospective, randomized trial of much less intense physical therapy was beneficial in 89 % of 28 patients, 8–17 years of age. The patients received physical therapy either once weekly or three times a week for 1 h. All patients received cognitive behavioral therapy for six 1-h sessions. Relapses were seen in 8/25 requiring more intense management; however, excellent functional status was reported in the long-term follow-up of 25 patients [70].

Maillard et al. reported a study of 23 patients with RSD treated either as inpatients or outpatients with a multidisciplinary approach including PT/OT, medications, and psychological intervention [71]. There was a direct correlation between the duration of symptoms and the initiation of therapy. Prompt intervention lessened the time of therapy required for improvement. There was no significant difference in recovery rates as a result of outpatient versus inpatient treatment. The average time to improvement was 4 weeks, and full recovery took an average of 25 weeks. Follow-up after 6 months showed full recovery in 58 %, while 26 % were improved but had residual symptoms and 15 % reported minimal recovery from symptoms.

In summary, patients with RSD/CRPS1 are usually female, experience lower extremity pain with edema, and develop vasomotor changes after minimal trauma. Some psychological aspects are present in many children. Response to physical therapy and cognitive behavioral intervention are beneficial in most patients. Early diagnosis and intervention offer an advantage for recovery. In most studies, relapses were common, but response to renewed therapy was good.

Chronic Widespread Musculoskeletal Pain/Fibromyalgia Syndrome

Case 4

A 16-year-old female with a known history of celiac disease presents to the office with a history of "everything hurts" for the past 6 months. She had a prolonged bout of "mono" prior to the onset of the pain that caused her to miss several weeks of school. The pain started in the shoulders and spread gradually to the back, elbows, hips, knees, and hands. She thinks her hands are swollen for an hour in the morning. In addition to the body pain, she has chronic headaches, abdominal pain with episodic diarrhea alternating with constipation, and extremely painful menses. She has a difficult time falling asleep, often taking up to 2 h with frequent arousals thereafter.

This has caused her to have severe fatigue. She is a straight A student, who is unable to participate in any activities but homework and chorus. In the past she was athletic but quit running cross country after the mononucleosis. Family history is positive for her mother who has fibromyalgia. She was treated with a graded aerobic exercise program and cognitive behavioral therapy that were gradually beneficial. She needed a lot of encouragement to do the exercise in the beginning, but it became easier over time.

Chronic WSP is common in children and adolescents, with increased prevalence with older age and female gender. Mikkelsson reported the development of WSP in 1,282 children. WSP developed in 7 % of 1,282 children at 1-year follow-up from baseline and 14 % at 4-year follow-up; however, only 3 % had the WSP at both 1 and 4 years [72]. Similarly the prevalence of pediatric fibromyalgia is reported in the same range of 1.2–6.2 % from different ethnic and geographic areas [7].

The juvenile fibromyalgia syndrome is characterized by chronic WSP in at least three areas for 3 months or longer. It is associated with chronic fatigue, sleep disturbance, and pain in other body parts such as headaches, chest pain, abdominal pain with irritable bowel syndrome, and numbness [8]. A feeling of swelling in the joints, especially the hands, is reported but the duration of the swollen feeling is usually short lived, lasting only an hour in the morning. When the patient is examined, there is no significant objective swelling present. The physical finding present in patients with fibromyalgia is the characteristic tender points in 18 specific anatomic sites (see Table 11.3).

The diagnosis of fibromyalgia was based on the Yunus criteria in earlier studies and the 1990 American College of Rheumatology (ACR) criteria in later studies; however, the 1990 ACR criteria have not been validated in children [8, 73]. The ACR 1990 criteria for the classification of fibromyalgia included widespread pain in combination with tenderness at 11 or more of the 18 specific tender point sites, present both above and below the waist. Yunus criteria require 5/18 tender points and at least three of the following ten features (1) chronic anxiety or tension, (2) fatigue, (3) poor sleep, (4) chronic headaches, (5) irritable bowel syndrome, (6) subjective soft tissue swelling, (7) numbness, (8) pain modulation by activities, (9) pain modulation by weather, and (10) pain modulation by anxiety/stress. Only 4/18 tender points were needed if 5/10 features were present. New preliminary diagnostic criteria for adults have been published that do not require physical or tender point examination [74].

Table 11.3 18 tender point examination

• Right and left low cervical C5–7
• Right and left occiput
• Right and left trapezius
• Right and left supraspinatus
• Right and left second rib at costochondral junctions
• Right and left lateral epicondyle at 2 cm distal to the epicondyle
• Right and left medial fat pad of the knee
• Right and left gluteal upper outer quadrant
• Right and left greater trochanter

The tender points have been systematically evaluated and are usually present in greater than 11/18 as demonstrated by Swain et al., who documented a mean of 15/18 and Buskila who found a mean of 12/18 [75, 76]. An additional physical finding that has been associated with fibromyalgia is joint hypermobility. Ting et al. found hypermobility in 48 % of 131 adolescents with fibromyalgia. The hypermobile patients had greater pain sensitivity and higher number of tender points compared to non-hypermobile patients with fibromyalgia [31].

Kashikar-Zuck et al. assessed the prevalence of mood, anxiety, and behavior disorders in 76 patients with fibromyalgia [77]. In this group, 67 % had at least one current and 71 % had a lifetime diagnosis of at least one psychiatric disorder. Current prevalence of anxiety was 57 %, of mood disorder was 22 %, and of behavioral disorder was 26 %. The authors concluded that the patients with fibromyalgia are predisposed to emotional difficulties, commonly in the form of anxiety, which is consistent with a stress-vulnerability model [77, 78]. Individuals in this model are predisposed to respond to stress by manifesting pain or psychological symptoms. Kashikar-Zuck et al. also found adolescents with

fibromyalgia to have greater internalizing symptoms (depression and anxiety) and externalizing symptoms (attention and conduct symptoms) compared to healthy peers [79]. In a comparison of patients with fibromyalgia to matched healthy peers, Kashikar-Zuck et al. found that adolescents with fibromyalgia had poorer physical functioning if the mother had a history of pain. The overall family functioning was poorer and exhibited conflicted families. They found that 24 % of the mothers reported having fibromyalgia [79]. The history should therefore include a query into the family functioning, maternal health history, especially the presence of a pain syndrome, and conflict in the family. These findings support the environmental impact on the development of fibromyalgia; however, there are also genetic variants of pain pathways that predispose to pain amplification. Research is evolving to partially explain the familial clustering of the idiopathic pain syndromes [10, 80, 81].

In a study of adolescent psychiatric inpatients, 52 % met criteria for diagnosis for fibromyalgia; thus providing insight to the complaints of chronic pain in this population [82]. In addition, there was a high incidence of prior physical or sexual abuse in this group of patients.

The prognosis of patients with fibromyalgia is often not favorable. Follow-up of 48 patients with fibromyalgia for an average duration of 3.67 years revealed that 62 % continued to have chronic widespread pain and significantly lower health status and physical functioning compared to healthy controls [83].

Treatment for fibromyalgia is multidisciplinary and requires a carefully shared, crafted plan with the patient and parents. This plan includes a tiered, graded aerobic and strengthening exercise program and cognitive behavioral therapy. Medications may be helpful but should not be the only treatment. These medications include amitriptyline, cyclobenzaprine, or fluoxetine; however, randomized controlled trials are lacking. Newer medications such as pregabalin or duloxetine which are potentially efficacious in adults have not been approved for use in adolescents. Cognitive Behavioral Therapy (Chap. 19) has been shown in a randomized clinical trial to be safe and effective for reducing functional disability and symptoms of depression in adolescents with fibromyalgia [84]. Patients in the CBT group received eight sessions and two booster sessions at later times. Patients are usually not readily accepting of psychological treatment but such therapy is empowering and gives patients self-management skills in the treatment and control of their symptoms. Additional psychological therapy may be required in the patient who has suffered either physical or sexual abuse.

Exercise therapy has been conducted in several randomized clinical trials for treatment of patients with fibromyalgia. The review by Busch et al. evaluates the benefits of exercise therapy which can be in the form of multiple types exercise including aquatic, aerobic, strengthening, yoga (Chap. 25), pilates, tai chi, etc. [85]. These studies support the recommendation for aerobic and strengthening exercise for treatment of fibromyalgia. Complementary and alternative therapies, such as tai chi, gi gong, massage therapy, and acupuncture (Chap. 23), have been utilized and should be entertained as potentially beneficial in motivated patients [86]. Care must be taken not to endorse "natural products" that have no evidence of efficacy or safety. The effectiveness of a comprehensive, multidisciplinary approach for patients with this complex biopsychosocial condition remains to be studied. A single therapeutic approach, however, does not seem to provide sustained benefit for these patients.

Questions

1. The most common cause of reflex sympathetic dystrophy in children is which one of the following:
   (a) Loss of a family member
   (b) School failure
   (c) Poorly defined trauma
   (d) Injury in a motor vehicle accident
2. Which of the following is contraindicated in the treatment of reflex sympathetic dystrophy:
   (a) Desensitization by rubbing with coarse towel
   (b) Immobilization

(c) Sympathetic ganglion blockade
(d) Aggressive mobilization

3. Which of the following physical findings suggests reflex sympathetic dystrophy:
   (a) Marked sensitivity to light touch
   (b) Insensitivity to pin prick
   (c) Erythematous, warm localized joint swelling
   (d) Hyperpigmentation of the skin around the hip girdle
4. Parents describe their child with growing pains with which of the following symptoms:
   (a) Morning stiffness and pain
   (b) Nighttime awakening with leg pains
   (c) Swelling and tenderness in the limb
   (d) Inability to ambulate after activity
5. The child with growing pains has which one of the following on physical examination
   (a) Limb length inequality
   (b) Atrophy of the calf muscles
   (c) No evidence of swelling or tenderness
   (d) Hyper-reflexia of patellar tendons
6. Which of the following symptoms is common in children with growing pains:
   (a) Headaches
   (b) Urticaria
   (c) Diarrhea
   (d) Low grade fever
7. Benign joint hypermobility may have joint swelling characterized by which of the following:
   (a) Warm, tender joint capsule
   (b) Joint held in 15° of flexion
   (c) Transient swelling after activity
   (d) Prolonged morning stiffness with pain
8. The patient with hypermobility is subject to developing which of the following conditions:
   (a) Anxiety
   (b) Fibromyalgia
   (c) Reflex neurovascular dystrophy
   (d) Bipolar disorder
   (e) Both (a) & (b)
9. Treatment for hypermobility is targeted at which of the following:
   (a) Treatment for depression
   (b) Strengthening and proprioception exercises
   (c) Immobilization of affected joints
   (d) Vitamin C repletion
10. Patients with fibromyalgia commonly complain of which of the following symptoms:
    (a) Chronic fatigue
    (b) Diarrhea and weight loss
    (c) Intermittent urticaria
    (d) Frequent aphthous lesions
11. Which of the following is contraindicated in the treatment of fibromyalgia?
    (a) Aerobic reconditioning
    (b) Cognitive behavioral therapy
    (c) Homebound virtual schooling
    (d) Tai chi and pilates
12. Which of the following patient profiles is typical of a patient with fibromyalgia?
    (a) Athletic male with a torn anterior cruciate ligament
    (b) A competitive swimmer with weight loss and amenorrhea
    (c) Tired, sedentary adolescent female with anxiety, migraines, and IBS
    (d) A girl with diarrhea, anemia, and migratory joint pain

Answers

1. (c): Poorly defined trauma is the most common antecedent historical recollection in the reflex sympathetic syndrome or CRPS I in children although often no event is recalled. Psychological trauma is possible however not the most common. Overt trauma is also possible but not the most common.
2. (b): Immobilization is contraindicated and actually aggravates the edema and pain in RSD. Conversely aggressive mobilization is the treatment of choice. Desensitization and sympathetic ganglion blockade are employed as treatments.
3. (a): The patients with RSD exhibit marked sensitivity to light touch or allodynia. This is characteristic of the pain syndrome. The swelling is typically diffuse and not localized to a joint per se. Color changes are common; vasomotor instability is characterized by pallor or erythema which wax and wane. Pigment changes are not characteristic.

4. (b): The child with growing pains often awakens at night with the pain in the lower extremities, muscles groups more commonly than joints. No objective signs are seen such as swelling; moreover stiffness in the morning is absent and refusal to ambulate is not typical.
5. (c): There are no objective signs on physical examination thus there is no evidence of swelling or tenderness. There is no evidence of previous inflammation such as limb length inequality or atrophy of muscles which is seen in longstanding juvenile idiopathic arthritis. Reflex changes are not characteristic of growing pains.
6. (a): Children with growing pains are subject to headaches which leads to the concept of more wide spread pain sensitivity. Objective signs of fever, rash, or diarrhea are lacking.
7. (c): Signs of inflammation such as joint warmth, flexion contracture, or morning stiffness are absent in benign hypermobility. The patients may have transient, short-lived swelling after activity; perhaps secondary to micro-trauma.
8. (e): Hypermobility is a common comorbidity in patients with fibromyalgia. Anxiety is described in patients with hypermobility; however, psychiatric disease such as bipolar disorder is not described. RSD is not a common comorbidity of hypermobility.
9. (b): The treatment of choice for hypermobility is strengthening and proprioception exercises to protect the lax joints from trauma and overuse of movement. Immobilization would ultimately weaken the joints from atrophy of muscle. Although some emotional contribution may amplify pain, depression is not a major feature of hypermobility. Vitamin D deficiency is seen in many chronic pain conditions; however, a treatment recommendation is not suggested unless a deficiency is identified.
10. (a): Chronic fatigue is one of the cardinal features of fibromylagia. Objective features of other inflammatory diseases are lacking, keeping in mind that patients with fibromyalgia may have a primary autoimmune disease and the fibromyalgia is a secondary feature of pain amplification. Diarrhea is a common feature of IBS; however, patients do not have significant weight loss. Frequent aphthous lesions suggest inflammatory bowel disease. Fibromyalgia is not associated with cutaneous manifestations.
11. (c): It is discouraged for patients with fibromyalgia to retreat from responsibility such as going to school. This maybe a difficult negotiation in some patients. Treatment is focused on reconditioning exercises and stress relief such as cognitive behavioral therapy and other exercise modalities such as pilates, yoga, and tai chi.
12. (c): Fibromyalgia has other features of centralized pain syndromes such as headaches, both migraine and tension type headaches, irritable bowel syndrome, anxiety, and fatigue. Although athletes may be subject to fibromyalgia, they are not the typical host for this condition. Diarrhea, weight loss, and migratory joint pain suggest inflammatory bowel disease.

Conclusion

The four most common chronic, recurrent musculoskeletal pain syndromes in children have a characteristic pattern of symptoms and signs that allow recognition of the diagnoses. These syndromes fit a biopsychosocial model with overlying psychological issues in many affected patients. Early recognition and treatment offer better outcomes and obviate excessive evaluation and potentially harmful treatments.

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Functional Symptoms in Infectious Disease: Reading into Titers

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Wendy A. Holz and Jana Shaw

Abstract

Most childhood infections are acute in nature and therefore less likely to result in emergence of functional symptoms typical of chronic conditions. However, human immunodeficiency virus infection (HIV) and Lyme disease, each for a different reason, remain exceptions. In the era of effective antiretroviral therapies, HIV has become a chronic illness. Children born with HIV face different stresses when compared to adolescents who acquire HIV through risky behavior. This chapter will provide a discussion of various mental health problems that accompany or mimic conditions associated with HIV infection that providers are likely to encounter when caring for children with HIV. Lyme disease is an acute illness with excellent prognosis when treated appropriately. Unfortunately, misleading information available on the Internet has fueled false beliefs about Lyme disease diagnosis, treatment, and prognosis. As a result, we have encountered a growing number of older children and adolescents with so-called chronic Lyme disease or post-Lyme disease syndrome. Because their definitions remain elusive, patients with subjective complaints often are diagnosed with Lyme disease. Proper knowledge of clinical manifestations of Lyme disease and interpretation of Lyme serology testing are paramount to prevention of unnecessary and misleading interventions. This chapter will focus on the challenges of Lyme disease diagnostics and treatment in patients presenting with functional symptoms.

Keywords

Lyme disease • HIV • Subjective symptoms • Anxiety • Depression • Fatigue • Pain • Adherence

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The management of certain infectious diseases can sometimes be complicated by the emergence of functional symptoms. The overlap between organic and functional symptoms can complicate the diagnostic workup and management.

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It is particularly important that the provider is aware of functional symptoms described in patients with HIV and Lyme disease. Unlike HIV, Lyme disease rarely results in chronic disease. The issue of chronic Lyme disease has a broader context because of patient-advocacy groups that promote months or years of antibiotic therapy for "chronic Lyme disease [1]." Moreover, chronic Lyme disease has become a common diagnosis for medically unexplained pain, neurocognitive symptoms, or fatigue, even when there is little or no evidence of previous Lyme infection.

This chapter provides brief introduction to both HIV and Lyme disease. The first part focuses on HIV, its clinical presentation, and common functional symptoms. The management of functional symptoms is usually handled by mental health specialists familiar with HIV patients. The first part concludes with several clinical HIV case scenarios and quizzes.

The second part briefly introduces Lyme disease, its clinical presentation, diagnosis, and management and details common functional symptoms ascribed to Lyme disease. Similarly to the first part, the second part offers a common case scenario and a quiz to offer a reader a better understanding of the spectrum of subjective complaints in patients with "chronic Lyme disease."

Key Organic Features of HIV

General Considerations

Human Immunodeficiency Virus/Acquired Immunodeficiency Syndrome (HIV/AIDS) is a disease of the human immune system caused by the HIV. The disease interferes with the immune system, which makes a person with AIDS much more likely to contract infections and cancers that do not typically affect people who have normal immune systems.

Causes and Incidence

HIV can be transmitted vertically (mother to child), through sexual activity, via contaminated blood products or IV drug abuse. Vertical HIV transmission occurs prior to birth, during the delivery or after birth. Transmission postnatally can occur due to the ingestion of breast milk infected with HIV [2]. There were 36,870 new HIV cases reported to the US Center for Disease Control in 2009 [3]. The rate of perinatally acquired HIV infection was highest among Black/African American children, at 9.3 per 100,000 live births in 2009 [4]. In 2010, there were an estimated 2,266 adolescents aged 13–19 years diagnosed with HIV infection in the USA [5].

With the implementation of prophylactic interventions, the incidence of vertical transmission has decreased significantly. The mother to child transmission rate of HIV in the USA is <2 % with appropriate prophylaxis. Most vertically transmitted infections in the USA are a result of not knowing the mothers' HIV status prior to delivery (40 %) [2]. While the rate of vertical transmission of HIV has decreased over the last decade, the number of infections acquired during adolescence is increasing with adolescents accounting for 50 % of all new HIV infections in the US [6]. In 2009, people aged 15–24 years accounted for 40 % of all new HIV infections [7]. HIV is also an enormous burden for families as well as those who are infected with HIV. The number of children and adolescents in the USA who are not HIV infected, but living with someone who is HIV infected is growing [6]. These children often stay with their parent regardless of the severity of the parent's HIV disease [6].

Clinical Findings

Acute HIV infection is defined as occurring during the time from exposure to HIV until the time that an HIV antibody test becomes positive [8]. If the HIV antibody test becomes positive, the patient is considered to have chronic HIV infection. Symptoms associated with acute HIV infection are often nonspecific and usually are thought to be the result of a viral illness. These include fever, chills, malaise, fatigue, swollen lymph nodes, sore throat, and myalgias [8].

Chronic HIV infection is divided into three classes based upon the CD4 cell counts. Patients with CD4 cell count >500/μl, 200–499/μl, and <200/μl are classified into A, B, and C category, respectively. The clinical symptoms range from no symptoms to generalized lymphadenopathy, oropharyngeal candidiasis (thrush), oral hairy leukoplakia, thrombocytopenia, anemia [8, 9], weight loss, HIV wasting syndrome (defined as an unintended and progressive weight loss), opportunistic infections, encephalopathy, and malignancy [9].

Diagnosis

The most commonly used test for HIV/AIDS is the HIV antibody test. A person generally develops HIV antibody 6–12 weeks after HIV infection occurs. Therefore, if an HIV antibody test is obtained prior to the production of antibodies, a false negative result will occur. For this reason, it is recommended that HIV antibody tests be performed 3 months after a potential HIV exposure [10]. HIV antibody tests are generally very accurate. If the screening test is positive, a Western blot assay is used for confirmation [10]. Fourth-generation tests look for both antibody and p24 antigen at the same time. The p24 antigen is an HIV protein that appears early after infection but declines with development of antibodies. This allows for earlier and more accurate HIV detection [10]. The polymerase chain reaction (PCR) test detects HIV genetic material and is used for babies born to HIV positive mothers because the babies retain their mothers' antibodies for up to 18 months, which makes an antibody test inaccurate. A PCR test is not usually used for testing in any other population as it is expensive and more complicated to interpret than the standard antibody tests [10].

Treatment

The goal of HIV treatment is to prevent or reverse the progression of clinical illness [11]. HIV infects and destroys immune cells (e.g., CD4). Effective therapy is evidenced by improvement of the signs and symptoms of HIV, a rising CD4 cell count, and a decreasing viral load.

Commonly Used Medications

- Antiretroviral medications used in a combination of at least three different forms from two different drug classes suppress viral replication by interfering with various steps of the replicative life cycle of HIV. Antibiotics are used for prophylaxis against opportunistic infections.
- Iron supplement for severe anemia.

Therapeutic Recommendations

- As most of HIV medications are processed through the liver, further insults to the liver should be avoided. Therefore, patients are counseled to avoid alcohol and tobacco.
- Live with a healthy lifestyle—exercise, balanced diet, adequate sleep.
- Condom use for sexual activity, to decrease transmission of HIV, to prevent contracting other sexually transmitted diseases or other strains of HIV, and to prevent pregnancy.

Prognosis

In general, if a patient presents early with HIV infection and adheres to the medication regimen he or she is expected to have a life expectancy similar to the general population.

Functional Conditions Associated with HIV

Prior to the availability of antiretroviral therapy, progressive HIV encephalopathy was well documented in children and included impaired brain growth, developmental delays or regression, and expressive language impairment to name few. The rate of encephalopathy was modified with

current antiretroviral therapy; nevertheless, residual behavioral, cognitive, neurologic, and scholastic impairments appear common among children with vertically transmitted HIV.

It is important to recognize that an adolescent who contracts HIV through behavioral factors will often have underlying psychiatric issues prior to the diagnosis of HIV. Studies show that adolescents with psychiatric disorders are at a greater risk of contracting HIV due to increased risk of unsafe sexual practices, impulsivity, self -destructive attitudes, cognitive immaturity, high rates of substance abuse, self injurious behavior, and sequelae of sexual abuse [12].

Within the pediatric and adolescent HIV population, 55 % meet the criteria for a psychiatric disorder [13]. Children with HIV infection also have additional risk factors for mental illness which include poverty, a disrupted home life, and a family history of mental illness, psychiatric disorders, substance abuse, and inadequate social support [14]. Children who live with HIV-infected parents are also at greater risk of developing psychosocial and psychiatric issues. Therefore it is very important that these children have a primary care provider who is familiar with their family history and is comfortable evaluating these patients for mental health issues. There is seldom access to the mental health care available to children and adolescents with HIV.

Anxiety

The most common mental health disorder in children and adolescents with HIV is anxiety, afflicting as many as 46% of 9–16-year-olds [15]. Anxiety in younger children often is related to issues surrounding a recent disclosure about their own or a family members HIV status or a change in the health status of a family member with HIV. Issues of their own or a loved one's health often create great concern within a child. A child with an HIV infected parent could be exposed to parental substance abuse, violence, and poor health of the parent all which can create anxiety. Adolescents deal with issues related to their HIV status disclosure and peer and sexual relationships. Parents of children with HIV have also shown tendencies to experience anxiety. Their triggers tend to be concern for their child's health and guilt associated with vertical transmission to their children.

Underlying medical conditions or HIV medications [16] can create symptoms that mimic anxiety symptoms such as pain, nausea, vomiting, tachycardia, abdominal pain, or shortness of breath. The list of organic causes that can cause symptoms that overlap with those of anxiety includes:

- CNS pathologies—HIV-related infections, neoplasms
- Systemic or metabolic illness—hypoxia, sepsis, electrolyte imbalance
- Endocrinopathies—thyroid disease, hypoglycemia, pheochromocytoma, Cushing's syndrome
- Respiratory conditions—pneumonia
- Cardiovascular conditions—arrhythmias, pulmonary embolus
- Substance intoxication/withdrawal—from alcohol, nicotine, caffeine, cocaine, and amphetamines
- Certain HIV medications can create anxiety-like symptoms

While organic causes can mimic anxiety symptoms, it is important to recognize that anxiety symptoms can also mimic organic causes of symptoms. For example, symptoms of anxiety that can overlap with HIV issues include insomnia and abdominal discomfort. A thorough history regarding the symptom is imperative to arrive at the correct diagnosis.

Behavioral Disorders

Nearly one-quarter of all children and adolescents with HIV meet the criteria for a behavioral disorder. Attention-Deficit Hyperactivity Disorder is the most prevalent (21 %) followed by conduct disorders (13 %) and oppositional-defiant disorders (11 %) [9, 11]. The etiology of behavioral disorders in HIV-infected children and adolescents is likely multifactorial including such factors as prenatal drug exposure, difficult family environment, age of parents, level of parental education, absence of parent, changes in

caregivers, nutrition, and poverty [17]. Children and adolescents with behavioral disorders are known to have greater difficulty in adhering to chronic medication regimen, thereby increasing risks of HIV-related illnesses and worsening prognosis.

Mood Disorders

Mood disorders affect 7 % of HIV-infected children and adolescents [15]. It is estimated that 6 % of children aged 9–17 years, experience some form of depression every year [17]. Poor adherence to medications and medical appointments were described in patients with HIV [18]. The stigma of HIV accounts for about 50 % of all depression in HIV infected adolescents [19]. Depression can also be caused by a number of other factors. Certain HIV medications, chemical changes in the brain, poor support at home, issues relating to keeping HIV status private, and concern over treatment failure are all common factors leading to depression [20]. Symptoms of depression that can overlap those of HIV are weight loss, insomnia, fatigue, and diminished ability to think or concentrate.

Substance Abuse Disorders

Substance abuse is a primary risk factor for HIV infection [21]. Although injection drug use is a risk factor of HIV transmission, it is generally not the cause of most HIV infections in adolescents as they tend to use oral and inhaled drugs more than injected substances [22]. Use of ecstasy (MDMA or 3,4-methylene dioxymethamphetamine) or amphetamines are on the rise among adolescents [23], which make them prone to engagement in risky sexual behaviors that put them or their sexual partner at risk for HIV.

It is well described that substance abuse and psychiatric orders often occur in tandem [24]. Illicit drugs can cause psychiatric disorders and they can also be used in an attempt to cure symptoms of a psychiatric disorder. Organic causes of behavioral changes in HIV infected infants and adolescents include HIV encephalopathy and CNS neoplasm or infection.

Post-traumatic Stress Disorder

The lifetime rates of post-traumatic stress disorder (PTSD) in an HIV infected person are at 10.4 % compared to 1.3–7.8 % of the general population [25]. Specific events that have been common in triggering the onset of PTSD in an HIV positive person are the diagnosis of HIV, the development of physical symptoms, initiation of antiretroviral treatment, and being close to a person who has an HIV-related death. In addition, emotional distress—fear, helplessness, or horror—has been associated with the development of post-traumatic stress [26]. While concerns are obvious for the HIV infected or affected adolescents, it is important to not overlook a child or an adolescent who might be experiencing PTSD as a result of a rape. Symptoms of PTSD that can overlap with organic issues of HIV include insomnia and an increase in HIV symptoms as a result of poor adherence to prescribed HIV therapy.

Adherence to Treatment

In HIV, adherence rates need to be at least at 95 % for the best therapeutic outcomes [27]. This is significantly higher than adherence rates required for most other chronic illnesses [27]. Non-adherence to a medication regimen in an HIV infected adolescent often results in viral mutations and resistance to the medication regimen prescribed.

There are several identified barriers to treatment. Complexity of the antiretroviral regimen can lead a patient to not take medications. Adherence has been shown to decrease with an increased pill burden. Children also have difficulty with the taste of the liquid medications. Children and adolescents who feel well often feel that they do not require medication [27]. Some adolescents are afraid that taking the medications will cause their diagnosis to be disclosed

inadvertently [24]. Expected side effects of antiretroviral medications, include nausea, vomiting, diarrhea, especially during the first weeks of therapy and can also lead to non-adherence. Also with adolescents, it is speculated that some non-adherence is related to normal developmental tasks of separation and individualization [28].

At times, it can be very difficult to differentiate between functional and organic symptoms of HIV. Patients with an unstable home environment and family history of mental health disorders are at higher risk for functional problems. It is very important to obtain a detailed psychosocial history including a symptom's triggers. If the history and/or laboratory testing suggest a functional problem, the patient should be referred to a mental health professional experienced with HIV. In addition, mental health screening should be part of routine HIV care even in clinically stable patients.

Case 1: Non-Adherence

The patient is a 20-year-old female diagnosed at 8 years of age with perinatally acquired HIV. Since 17 years of age, her adherence became intermittent. HIV was not discussed in the home at all. No one outside her mother and father knew of her HIV status. Her mother reported that she did not even discuss her HIV status with her doctor. The patient requested not to be told her CD4 counts and viral loads so as not to be reminded of her disease. The patient was offered mental health counseling but she did not keep her appointments. In her first year of college away from home, the patient made new friends to whom she disclosed her HIV. She reported that her friends were very supportive and accepting of her regardless of her HIV diagnosis. She was taking her medication as prescribed. After a year, she returned back home to her parents and went to a local community college. Her viral load began to rise and her CD4 count began to decline. During her routine follow-up visits, she would not make eye contact, often cry, and pull her eyelashes. She denied headache, fever, photophobia, paresthesia, or numbness. Her neurological exam was normal. She stated that every time she tried to take her medications she would be reminded of her HIV. She admitted that she did not feel that she had support from her family as she did with her friends at her first college.

Questions

1. The patient's symptoms can most likely be associated with which mental health issue?
   (a) Anxiety
   (b) Depression
   (c) Bipolar disorder
   (d) Normal, developmentally associated non-adherence
   (e) Drug abuse
2. Which one of the following factors is most likely at the root of recent deterioration in her mental health status?
   (a) Her family inability to discuss HIV and answer any questions that may have surfaced after the initial conversation.
   (b) She misses her college friends.
   (c) It is a common side-effect of her antiretroviral medications.
   (d) She is disappointed in her performance in college.
   (e) It is a manifestation of an opportunistic CNS infection.
3. What would be the next best treatment approach with this patient?
   (a) Offer adherence counseling.
   (b) Continue to encourage and prescribe antiretroviral therapy.
   (c) Offer treatment for her underlying depression with both medication and counseling.
   (d) Encourage open discussion at home.
   (e) All of the above.

Answers

1. (b): This patient shows signs of clinical depression. The symptom of trichotillomania suggests anxiety as well. Referral to a mental health specialist is most appropriate at this time. Drug abuse is unlikely given absence of other symptoms.
2. (a): This patient has been brought up to believe that her HIV is a big secret. While it may not be appropriate to disclose to people she does not know well, not allowing her to ask questions or discuss her fears or concerns with her

family, likely has worsened her depression and anxiety. CNS infection is unlikely given absence of neurological symptoms.

3. (e): This patient will not be able to adhere to antiretroviral therapy unless her depression is treated as well. Since her HIV is now poorly controlled, it is important to address all areas to facilitate her adherence.

Case 2: Difficulty Swallowing Pills

The patient is a 17-year-old female, perinatally infected with HIV. Her mother had a history of severe depression and schizophrenia.

Initially, the patient did well but as she grew older she started skipping her medications. Most recently, she stated that she had not been taking her medications because she felt that they had become lodged in her throat. She denied any problem with swallowing food or drinks.

Given her poor adherence, mental health counseling and psychotherapy were initiated. She was diagnosed with anxiety and mild depression and started on pharmacotherapy. She said that she was able to tolerate those medications because the pills were smaller but she refused her HIV medications. An adherence counselor was consulted, and she started hypnosis therapy. Because her viral load and CD4 counts deteriorated, her regimen was changed to liquid formulation. She did not like the taste of the liquid medications and ultimately stopped taking those as well.

As a result of her non-adherence, she developed HIV wasting syndrome. A gastrostomy tube placement was complicated by a gastrointestinal bleed. She underwent an upper endoscopy that revealed an esophageal stricture.

Questions

1. Did this patient have signs and symptoms of an esophageal stricture that should have been addressed earlier?
   (a) Yes. HIV alone is a risk factor for esophageal strictures.
   (b) Yes. The fact that she was able to swallow small pills but struggled with larger ones should have prompted earlier diagnostic study regardless of her ability to swallow food.
   (c) No. The patient had no risk factors.
   (d) No. This patient had anxiety that could easily explain her non-adherence.
   (e) No. She even refused liquid medication suggesting a functional problem.
2. What would be an appropriate plan of care for the patient at this time?
   (a) Dilate the stricture and remove the gastrostomy tube as soon as the stricture is found to be resolved.
   (b) Leave the stricture alone. A gastrostomy tube is in place and use it for medications and enteral nutrition.
   (c) Dilate the stricture and keep her on liquid formulation of medications.
   (d) Keep the gastrostomy tube in place until she can tolerate oral medications.
   (e) Dilate the stricture, keep the gastrostomy tube in place until she can tolerate oral medications and, start enteral nutrition to reach optimal weight and arrange for counseling to address mental health needs.

Answers

1. (b): Gastro-esophageal reflux and caustic ingestion are the most common risk factors for an esophageal stricture in children. HIV alone is not a risk factor. However, patients with poorly controlled HIV are at high risk of esophagitis, due to opportunistic infections, which can result in stricture. Although this patient had a history of non-adherence, anxiety, and depression, the fact that she was able to swallow smaller pills point strongly to esophageal pathology, regardless of her ability to swallow food. The cause of her stricture remained unclear.
2. (e): Stabilizing her HIV infection and optimizing her weight are most likely achieved by dilating the stricture, keeping the gastrostomy tube in place, and starting her on enteral nutrition. While the ultimate goal is to get the patient back on the pill form of her medications, her recovery can be expected to be a long process and unlikely to be achieved until her HIV and mental health are brought under the control.

Case 3: Decreased Appetite, Nausea, and Abdominal Pain

A 16-year-old female is referred to the clinic as she was recently diagnosed with HIV. She reports that her mother died in a car accident a few years ago and since then she feels that she has no one who understands her. She has been using marijuana, alcohol, and cigarettes. She has had unprotected sex with many sexual partners. Recently, she developed decreased appetite, nausea, and abdominal pain. She denies weight loss. She has not disclosed her HIV status to her family as she is afraid that they will reject her. She feels that she has no one to talk to about her HIV. Her physical exam shows abdominal tenderness in all four quadrants, but is otherwise unremarkable. Her initial laboratory evaluation including complete blood count, comprehensive metabolic panel, amylase, lipase, urine analysis, and abdominal X ray are all within normal limits. However, her CD4 count is low and viral load is high.

Questions

1. What is the possible explanation for her abdominal pain and nausea?
   (a) Mesenteric lymphadenitis.
   (b) Sexually transmitted disease
   (c) Anxiety
   (d) Pregnancy
   (e) All of the above
2. What is the next step in patient's management?
   (a) Admit her to a drug and alcohol treatment center.
   (b) Refer to mental health and perform pelvic exam, to rule out sexually transmitted diseases, and add pregnancy test.
   (c) Have her evaluated by a mental health specialist as she clearly has anxiety and depression related to her recent diagnosis.
   (d) Prescribe an antiemetic and follow-up in 1 week.
   (e) Admit her to the hospital, start antacids, and initiate antiretroviral medications once her abdominal pain resolves.
3. When should antiretroviral therapy be initiated?
   (a) Now, since her CD4 count is low and her viral load is high. You should not wait since she will be at a risk for opportunistic infections
   (b) Wait until she is no longer feeling nausea and abdominal pain and then start.
   (c) Discuss the need to start medications, arrange for mental health specialist counseling to discuss importance of adherence, and address substance abuse.
   (d) Tell the patient that she needs to stop drinking prior to giving her the antiretroviral medications in order to minimize hepatoxicity.
   (e) Do not treat since patient is likely to be non-adherent to medications and increase her risk of resistant HIV infection.

Answers

1. (e): This patient's presentation illustrates how difficult it can be to differentiate functional symptoms from organic condition. Given the patient's risky behavior, an organic cause has to be first ruled out. Diagnosis of a functional disorder is supported by the absence of laboratory abnormalities, diffuse nature of her abdominal pain, along with the stress related to her recent HIV diagnosis. The patient should be referred to a mental health specialist for further evaluation.
2. (b): Since the preliminary evaluation is non-revealing, pregnancy and sexually transmitted infections should be ruled out. In addition, patient should be referred to a mental health specialist even if her abdominal pain is organic in nature since the patient is expressing difficulty with disclosing her new diagnosis. In addition, her substance abuse needs to be addressed in order for her to be successful with antiretroviral therapy when that is initiated.
3. (c): Adherence to antiretroviral therapy is known to be affected by mental health issues and substance abuse. If possible, these issues should be addressed prior to initiation of therapy. In some situations, it may not be possible to wait due to the patient's medical condition

resulting from poorly controlled HIV. In those cases, it is important to work together as a multidisciplinary team to address goals of both the practitioners and the patient.

Key Organic Features of Lyme disease

General Considerations

Regardless of the clinical presentation, most patients with Lyme disease have resolution of their clinical symptoms when treated with appropriate antimicrobials. Persistent or recurrent symptoms after therapy are often due to functional problems or re-infection [29]. Extensive publicity has resulted in a degree of anxiety about Lyme disease that is out of proportion to its actual morbidity. Many physicians who promote chronicity and morbidity of Lyme disease belong to the International Lyme and Associated Diseases Society (ILADS) or label themselves as "Lyme-literate" doctors. Lyme-literate doctors propagate the idea of "persistent infection" within immune and neural cells and immune evasion of the bacteria and propose long, preferably intravenous therapy. They advertise their theories on the internet, in lay publications, and in the political arena [1, 30]. There is no scientific evidence to support these hypotheses. Unfortunately, persistent misconceptions result in unnecessary patient anxiety, cost, and side effects of prolonged antibiotic use. Scientific evidence, as documented by well-controlled NIH trials, argues against all of these false concepts.

Causes and Epidemiology

Lyme disease is an infection caused by a bacterial spirochete, *Borrelia burgdorferi*, which is transmitted by the bite of the tick species *Ixodes scapularis* and *Ixodes pacificus*. In 2010, 96 % of Lyme disease cases were reported from 13 states in parts of the mid-Atlantic and in parts of Minnesota and Wisconsin [31]. The incidence was reported to range between 9.3 and 84.6 per 100,000 persons in these prevalent areas in 2011 [32]. The incidence of Lyme disease among children aged 5–10 years (23 per 100,000 persons) is twice that of among adolescents and adults aged 20–35 years (8–12 per 100,000 persons) [31].

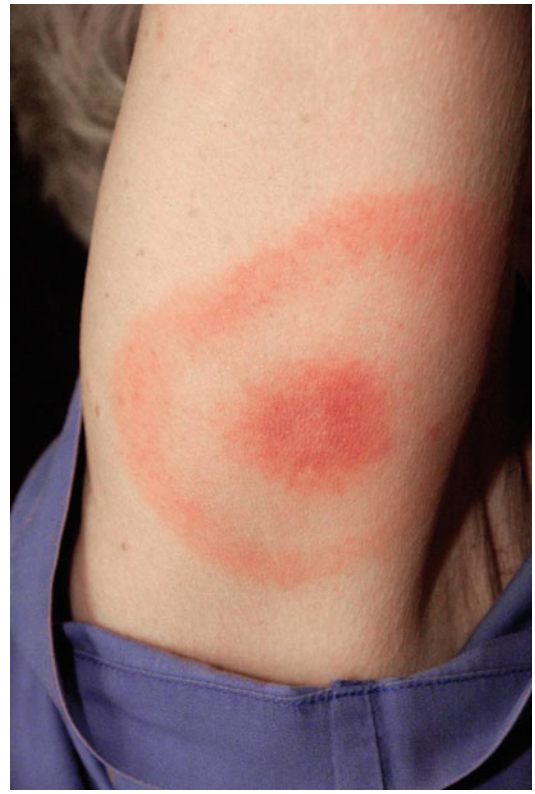

Fig. 12.1 Classic erythema migrans rash (Reprinted from Centers for Disease Control and Prevention (CDC). Public Health Image Library (PHIL): Photographs, Illustrations, Multimedia. ID#9875. http://phil.cdc.gov/phil/details.asp?pid=9875. Courtesy of Dr. James Gathany)

Clinical Manifestations

Early symptoms include fever, headache, fatigue, and a characteristic skin rash called erythema migrans (EM) (Fig. 12.1). If left untreated, infection can spread to the joints, heart, and nervous system [33]. The classic EM occurs at the site of a tick bite, 3–30 days (typically within 7–14 days) after the bite. Common manifestations of early disseminated Lyme disease in the USA

include multiple EMs, cranial nerve paralysis, and infection of the membranes covering the brain. Systemic symptoms such as fever, myalgia, arthralgia, headache, and fatigue are also common in this stage of Lyme disease. Carditis, which usually manifests as a prolonged PR interval or complete heart block are rare manifestations of early disseminated disease. Late manifestations of Lyme disease in children include arthritis and rarely infection of the brain [33].

Diagnosis

EM is diagnostic for Lyme disease but extracutaneous manifestations require antibody testing to confirm the diagnosis [33]. Only a minority of patients with single EM will have a positive test result because the rash usually develops before the antibodies are detectable. However, any other clinical manifestions of Lyme disease are accompanied by positive antibodies. The confirmation of Lyme disease by the laboratory usually rests on the demonstration of antibodies to *B. burgdorferi* in the patient's serum. Diagnostic testing should be performed in laboratories with excellent quality-control procedures for confirmation of extracutaneous Lyme disease. A two-step procedure implements first, a sensitive screening test, such as an enzyme-linked immunosorbent assay (ELISA) and, if that result is positive or equivocal, a Western immunoblot to confirm the result. The interpretation of those tests is straightforward [33]. Patients who test positive by screening ELISA will automatically undergo Western blot testing to rule out false positive results. Antibody tests for Lyme disease should not be used as screening tests in patients with chronic subjective symptoms alone since Lyme disease rarely, if ever proves to be the cause of the patient's symptoms [34]. False diagnosis of Lyme disease results from multiple factors and includes: provider who is not familiar with the interpretation of Lyme disease testing (positive screen test and negative Western blot means that the test is falsely positive), use of laboratory testing with poor quality controls, persistence of Lyme titers is viewed by some providers as persistence of infection, etc.

Treatment

Most patients with Lyme disease can be treated successfully with a few weeks of antibiotics. Early disseminated disease and arthritis are best treated orally with doxycycline or amoxicillin [35]. Additional treatment with nonsteroidal anti-inflammatory drugs may also provide symptomatic relief. Intravenous therapy with ceftriaxone is often used for Lyme meningitis, myocarditis, and heart block in symptomatic patients requiring hospitalization [33].

The long-term prognosis for individuals who are treated with appropriate antimicrobial therapy for Lyme disease, regardless of the stage of the disease, is excellent [33, 34, 36]. Antibody testing should not be used as a guide to length of treatment since they are expected to remain elevated for months to years after successful treatment. The elevated antibody levels are a hallmark of immunity, rather than a sign of an ongoing infection.

Functional Conditions Associated with Lyme Disease

In general, Lyme disease is an acute illness and objective long-term complications are uncommon. Therefore, functional problems associated with other chronic medical conditions are rare in children with true Lyme disease. In fact, children almost never have nonspecific symptoms as the only manifestation of Lyme disease [37]. Many different psychiatric symptoms have been reported in adults in association with Lyme disease including depression, mania, delirium, dementia, psychosis, obsessions or compulsions, panic attacks, catatonia, and personality change [38, 39]; however, causality by Lyme disease has never been clearly established for most of these symptoms. In addition, a diagnosis of Lyme disease should not be made on the basis of positive serologic testing alone. In many of the case reports, positive serology (with or without a classic history of Lyme disease) may be coincidental. Unfortunately, a number of patients with subjective symptoms such as fatigue, myalgia, or poor mental concentration with falsely positive Lyme

screening tests end up on repeat oral and prolonged parenteral courses of antibiotics, unnecessarily [34, 36]. Those patients are misinformed by "Lyme-literate" physicians and treated for Lyme disease when in fact their symptoms are a result of underlying psychosocial stresses or an unrecognized mental health condition.

Post-Lyme Disease Syndromes

According to the Infectious Disease Society of America guidelines there is no well-accepted definition of post-Lyme disease syndrome (PLDS) [35]. This has contributed to confusion and controversy and to a lack of firm data on its incidence, prevalence, and pathogenesis. Patients with long term post-treatment symptoms that are severe enough to be disabling are often labeled as PLDS. Although symptoms such as pain, fatigue, and difficulty with daily activities are common among patients treated for Lyme disease at some point in their lives, the frequency of such symptoms is similar in individuals who have never had Lyme disease [40]. The uncertainty whether PLDS even exists can lead to conflict between patient and healthcare provider over the diagnosis and treatment.

It is highly unlikely that PLDS is a consequence of occult infection of the central nervous system. This conclusion is based on evidence such as the absence of inflammation in the cerebrospinal fluid [41, 42] and negative results of both cultures and PCR assays for *B. burgdorferi* in the cerebrospinal fluid [37, 38]. Further, there is an absence of structural abnormalities of the brain parenchyma [43] and normal neurologic function, with no effect of antibiotic therapy on cognitive function [41, 44].

Chronic Lyme Disease

The term chronic Lyme disease is similarly poorly defined but widely used. Often, the term chronic Lyme disease is used as a diagnosis for patients with persistent pain, fatigue, or neurocognitive complaints, without clinical evidence of previous acute Lyme infection [1, 35] This viewpoint is fueled by a belief that Lyme disease causes serious disabling subjective symptoms even in the absence of objective signs of the disease, that diagnostic tests for extracutaneous manifestations of Lyme disease are falsely negative, and that prolonged treatment with antibiotics is necessary to alleviate the symptoms. The differential diagnosis of chronic Lyme disease includes a broad range of mental health conditions such as anxiety, depression, conversion disorders, chronic fatigue syndrome, fibromyalgia, confusional states, and acute memory disorders. Patients with objective clinical findings such as nerve paralysis, arthritis, or heart block should be promptly referred to a healthcare provider for further diagnostic evaluation and proper treatment.

Treatment of Patients with Post-Lyme Disease Syndromes and Chronic Lyme Disease

There is no convincing biologic evidence for the existence of symptomatic chronic Lyme infection among patients after receipt of recommended treatment. Antibiotic therapy has not proven to be useful and is not recommended for patients with chronic (>6 months) subjective symptoms after completion of recommended treatment regimens [35]. Attention has therefore turned to other explanations of chronic symptoms after Lyme disease and alternative therapeutic approaches to management. Many patients with PLDS fear that their symptoms are indicative of a chronic infection that may cause neurological damage. These concerns should be openly addressed and the patients reassured. There is no substitute for sympathetic listening and explanation [42]. The scientific evidence against the concept of chronic Lyme disease should be discussed and the patient should be advised about the risks of unnecessary antibiotic therapy. The patient should be thoroughly evaluated for medical conditions that could explain the symptoms and undergo a careful psychosocial evaluation. For example, we use the "Heads First" checklist (Fig. 12.2) [45].

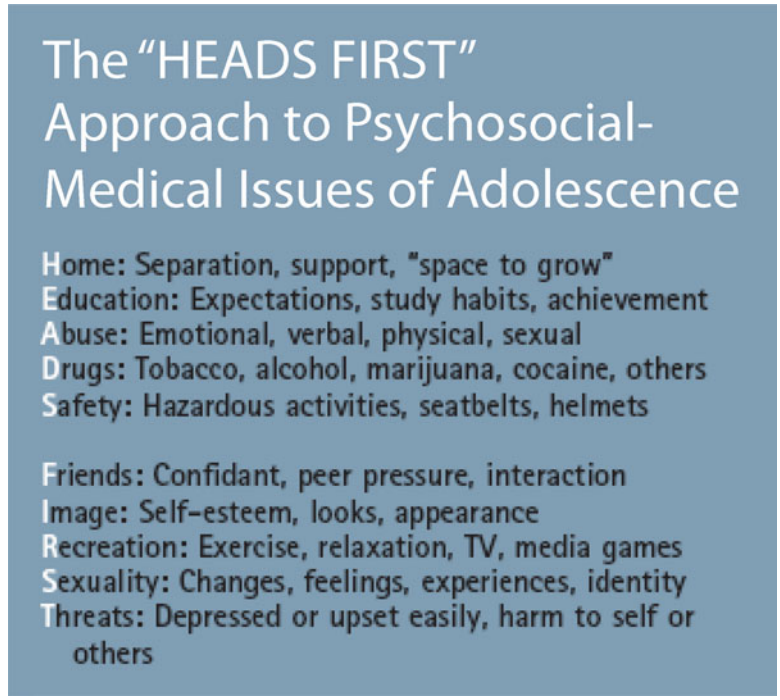

Fig. 12.2 The "HEAD FIRST" (Reprinted from Cavanaugh [45] with permission from American Academy of Pediatrics)

In many instances, the information obtained during the psychosocial interview is key to elucidating the cause of unexplained fatigue or pain in a teenager. If a diagnosis for which there is a specific treatment cannot be made, the goal should be to provide emotional support and management of pain, fatigue, or other symptoms as required [45–48]. If patient is accepting of a nonorganic cause as an explanation for his or her symptoms, a referral to an adolescent specialist or psychiatrist for psychological therapy could improve the patient's physical and emotional well being.

Differentiation of Functional from Organic Symptoms in Lyme Disease

Neuroborreliosis (brain disease as a result of a Lyme infection) is characterized by subjective AND objective findings of stiff neck, palsy, sensation, tingling, or weakness and positive Lyme serology. Patients with so-called chronic Lyme disease lack objective clinical and laboratory findings consistent with Lyme disease.

The differential diagnosis of neuroborreliosis in a patient presenting with fatigue, headache, depression, or impaired cognition includes fibromyalgia, chronic fatigue syndrome, other infections, somatoform disorders, depression, autoimmune diseases, and multiple sclerosis [38, 39, 49].

Two-step diagnostic Lyme testing in a patient with clinical symptoms and objective findings consistent with Lyme disease is the gold standard for diagnosis. On the other hand, subjective findings consisting of headaches, fatigue, neck pain, poor appetite, sleeping disorder, affected school performance and daily activities, memory, or concentration problems in a patient with negative Lyme testing argue strongly against Lyme disease.

Therapeutic Approaches

Chronic Lyme disease has become a common diagnosis for medically unexplained pain, neurocognitive or fatigue symptoms and requires an appreciation of its complex psychopathogenesis.

Unfortunately, attempts to deny the diagnosis or to offer an alternative explanation and treatment often are dismissed by the patient with anger and animosity. Interviewing patients about their worries, their ways of soothing, and what they think their symptoms represent will enhance diagnosis and treatment of each patient. This path of inquiry implicitly suggests there is more to the symptom than "just" the organic component and that the patients contain the key to what the symptoms mean. The path to self-awareness begins with noting the role of the unconscious in our health and wellbeing, and this can begin during the physical exam through sensitive inquiry. Self-awareness is what is often most underdeveloped in those prone to functional symptoms. The provider can then use their authority as an expert to reassure the patient that worry about symptoms exacerbates the symptom. This affords a more fluid transition to referring the patient for treatment of the anxiety.

The most effective current treatment for medically unexplained symptoms is a combination of therapeutic approaches tailored to the individual patient [50]. Most patients are in need of psychotherapy, relaxation therapy such as with use of hypnosis (Chap. 21), graded exercise therapy, cognitive behavioral therapy (Chap. 19), and physiotherapy under medical supervision. Psychotherapy with supportive counseling and family therapy sessions frequently are beneficial. Pain management with acetaminophen or nonsteroidal anti-inflammatory medications such as ibuprofen should be prescribed as indicated for headache, arthralgia, myalgia, and other such discomfort. Symptoms of depression, with insomnia or nighttime restlessness, can be treated with sedating antidepressants. Symptoms of depression with hypersomnia or psychomotor retardation may respond to selective serotonin reuptake inhibitors, which tend to have a more stimulating effect in most individuals. If fatigue is the main symptom, treatment is supportive because there is no known cure. Patients should be encouraged to gradually return to school as soon as possible to avoid the vicious cycle of fatigue, depression, social isolation, and school phobia [45].

Case

A 15-year-old girl comes to your office with 6 months history of dizzy spells that were associated with abnormal twitching movements and syncope. Her twitching movements resolved during sleep. She had normal evaluation by a cardiologist and neurologist. She was then seen by "Lyme specialist" who diagnosed her with Lyme disease in spite of her several negative Lyme serologies. She was placed on doxycycline for 3 weeks but her symptoms did not improve. Hence, she was started on oral rifampin then cefuroxime and subsequently had a long-term intravenous line placed and was treated with intravenous ceftriaxone for 3 months. Her twitching and frequency of syncope worsened regardless of ceftriaxone treatment and she was hospitalized for further evaluation. Repeat neurology and cardiology examinations were normal. The patient remembers that her symptoms worsened with stresses and that she became very upset the day of admission because she learned that she would be unable to join the varsity soccer team as a result of her syncope.

Questions

1. The most likely diagnosis for this patient's twitching movements and syncope is:
   (a) Seizure disorder
   (b) Vasovagal syncope
   (c) Conversion disorder
   (d) Anxiety disorder
   (e) Partially treated chronic neuroborreliosis
2. Which statement is correct regarding prolonged and repeated antibiotic courses in the treatment of chronic Lyme disease?
   (a) They have not been helpful in alleviating patients' subjective symptoms
   (b) They have been associated with serious side effects
   (c) They delay appropriate diagnostic evaluation
   (d) They are promoted by "Lyme-literate" doctors
   (e) All of the above
3. Which of the following is true regarding chronic subjective symptoms such as pain,

neurocognitive symptoms, or fatigue in the absence of objective findings and negative Lyme disease testing?

(a) Lyme testing should be repeated in specialized laboratories
(b) Patients with such symptoms should be treated with antibiotics regardless of the Lyme disease testing
(c) Such symptoms are common among children
(d) Suggest chronic neuroborreliosis and should be confirmed by spinal tap
(e) None of the above

Answers

1. (d): The most likely explanation is anxiety. The patient's symptoms were aggravated by stress. Inpatient psychiatry evaluation revealed an underlying anxiety. The patient's antibiotic treatment was discontinued and she was discharged home under the care of a psychiatrist.

   Chronic seizure disorder would likely be detected by EEG and not resolve during sleep. A vasovagal syncope is unlikely given the lack of a prodrome and a characteristic trigger. Chronic neuroborreliosis is unlikely given lack of objective findings, negative Lyme serology, and sufficient prior treatment with antibiotics.
2. (e): Patients with diagnosis of chronic Lyme disease require emotional and psychological support and avoidance of additional antibiotic courses. Alerting patients regarding internet sites with false and unscientific information about Lyme disease is very important. The following internet link provides very helpful information and videos about Lyme disease: http://www.cdc.gov/lyme/, http://www.idsociety.org/lyme/, http://www.idsociety.org/Chronic_Lyme_Video/
3. (e): Persistence of symptoms in patients appropriately treated for Lyme disease is commonly explained by other illness than chronic Lyme infection. Patients with chronic subjective symptoms and absence of objective findings should not be tested further or receive additional antibiotics. Instead, a collaborative approach between physician and patient is crucial to the goals of palliation of symptoms and rehabilitation, and the patient should be encouraged to take an active role in the treatment process.

Conclusions

Functional issues arise during HIV infection and require early recognition by the provider to optimize treatment. Mental health specialists familiar with HIV are ideal for initial and if available for subsequent mental health care.

Many different subjective symptoms have been reported to be associated with Lyme disease. However, a causal relationship with Lyme disease has not been clearly established for most of these symptoms. In the absence of objective clinical findings or laboratory abnormalities, the provider should perform a careful psychosocial evaluation and consider mental health services. Most patients with chronic Lyme disease are reluctant to accept a possible functional disorder as the cause of their symptoms. Therefore, it is critical to establish compassionate, respectful and collaborative relationship with the patient to optimize care and avoid needless antibiotic therapy.

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Functional Symptoms in Medical Genetics

13

Robert Roger Lebel

Abstract

Genetic disorders create significant pathophysiologic states that are the underpinnings of adverse experiences for affected individuals. Parents and health professionals face the challenge of distinguishing which reports of adverse experience call for medical intervention and which are best approached with empathy and environmental or psychological alterations. Parents and health professionals will seldom accomplish much by denying the existence of a symptom reported by a child; if the symptom does not connote a dangerous pathophysiologic state, that happy fact does not make it less real as an experience confronting the child. All the more difficult is the challenge when it is encased in a known foundational biological state resulting from a genetic disorder, and still further if it is also involving a reporter with intellectual disability. This does not make the challenge insoluble. Examples are discussed from the lives of persons affected by Down syndrome, velo-cardio-facial syndrome, and phenylketonuria. All three scenarios represent composite experience with persons affected by the three conditions in question.

Keywords

Genetic disease • Aneuploidy • Autosomal recessive inheritance • Chromosome deletion • Down syndrome • Velo-cardio-facial syndrome • Phenylketonuria

Background

In the long-standing "nature vs. nurture" dialectic, there is a persistent temptation to accept a binary assumption that deludes one into claiming pre-eminence of one over the other. Such an intellectual position amounts to blind faith and makes no contribution to progress in understanding actual human phenomena. For example, "intelligence"

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is not a product of nature or nurture, but a profoundly complex characteristic which is the product of innumerable components (neural circuitry, nutrition, familial influences, experiences, biochemical substrates, accidents, etc.) It is not a competition between nature and nurture but the outcome of their delicate and intimate (but usually not prescribed or directed) interactions.

Genetics is quintessentially the study of biological underpinnings for health and disease. It is understandable that both scientists and lay persons would perceive the articulation of a genetic etiology for some health problem as defining its source, its extent, and its very limited opportunity for intervention outside of direct molecular therapy for what is essentially a molecular problem. Yielding to the seductive opportunity to simplify matters does not serve the patient well, nor does it advertise wisdom and subtlety in the practitioner. Usually it will not be the genetic diagnosis but the symptoms themselves that dictate interventions. We shall explore some contributions of functional symptoms to the phenomenology of three genetic disorders.

Diagnosis

Diagnosis of a genetic disease is accomplished by the marshaling of individual and family history along with physical examination and targeted studies. The studies include radiographs and other imaging to provide information about anatomic structures not accessible to the hands and eyes of the examiner, as well as laboratory studies to investigate the microscopic and the submicroscopic components of heredity vehicles.

It is difficult to overstate the importance of a final precise genetic diagnosis. Without one, families are fearful of uncertain outcomes and of recurrences, and they are also vulnerable to unscientific claims of nonspecific therapeutic promise. In the first place, a final diagnosis means an end to the "diagnostic odyssey" upon which one enters when there is an unexplained health problem; concluding that journey is a benefit per se even if the final diagnosis has unpleasant implications, because it also concludes expensive and/or uncomfortable diagnostic efforts. Secondly, it provides to the patient, the family, and healthcare team the opportunity to read appropriate literature in order to have clarity as to prognosis and best therapeutic practices. Third, it provides to all involved an appropriate set of expectations for prognosis (likely or unlikely complications, survival, quality of life, etc.) Fourth, it allows for a meaningful discussion of recurrence risk in future members of the same family (which may be less than 1 %, or may be as high as 50 %). Finally, it allows accurate assessment of what measures (if any) are available to prevent or avoid the birth of another affected family member.

The prognostic and therapeutic narrative is, however, laden with dangers of a fatalism founded on the reductionist notion that genes are equivalent to destiny. There are many settings in which powerful interventions run contrary to that assumption. Many of the problems associated with functional symptoms of genetic disorders emerge from capitulation to that fatalism. Environmental events are every bit as important to the health and illness of persons with genetic disorders as for others.

Case Studies

Down Syndrome

This condition is associated with a discrete aneuploid state (unbalanced chromosome constitution, in this case the presence of three rather than the normal two copies of chromosome 21). Ordinarily an individual receives exactly 23 chromosomes from each parent. Most extra or missing chromosomes have such severe consequence that they precipitate early miscarriage. But trisomy 21 often allows for continuation of pregnancy. At birth, half of babies with this condition, which goes by the clinical name "Down syndrome (DS)," have cardiovascular malformations. Many have other anomalies, including a characteristic facial gestalt that usually makes

Fig. 13.1 A boy with Down syndrome

them readily recognizable to an experienced pediatrician or pediatric nurse (Fig. 13.1). Affected persons have developmental milestone delays but their severity cannot be predicted at birth and remains to emerge in the affected person's developmental future [1].

Persons affected by Down syndrome usually learn to walk, only later than children with typical chromosome constitution. They usually learn to talk, only later and with more limited final vocabulary. Their level of intellectual disability is quite variable, but it is almost always impossible for them to engage in abstract thinking, so that while they might learn simple addition and subtraction they cannot accomplish higher mathematics. They are almost never able to obtain a license to drive an automobile or to hold down a job complex enough to allow financial independence. They require supervision from someone with typical intellectual skills to manage, for example, a checking account. They are prone to hypothyroidism at any time of life, and in middle age they are prone to early-onset dementia [2].

The mother of an adolescent with Down syndrome once told this author that she did not have much trouble with him when he was acting intellectually disabled, but rather when he was acting like a normal teenager. Her irony was intentional, and the topic of conversation was rebelliousness and stubbornness in the face of the typical transitions of adolescence. This period in the life of a person affected by Down syndrome offers us two examples of functional symptoms.

(a) Experiencing otherness. As a child with Down syndrome observes peers at school who are obtaining drivers' licenses, or planning for college away from home, s/he is painfully aware of being unable to obtain equivalent goals. Parental fears upon learning of the infant's diagnosis become realized in these frustrations of perfectly normal aspirations. A thoughtful and provocative discussion of this problem in the context of self-awareness can be found in "Differences in Common," by Marilyn Trainer [2] (pp. 139–144).

Anyone, with any level of cognitive ability, may have moments of being less than patient with life's limitations. Anyone might then behave petulantly or unconstructively. More so the person whose self-awareness is also limited and the unarticulated sense of being "different" can manifest with sullenness that can form a pattern of stunted progress. So, the problem is not a specific feature or manifestation of Down syndrome, but rather something in the spectrum of behavior for any person whose life is not following his/her expectations. Intellectual disability accentuates and complicates it (Chap. 14).

(b) Somatization. Most medical literature about somatization involves persons of typical intellectual ability, but the problem can be complicated by diminished capacity. In her book cited above, Marilyn Trainer recounts an episode of sudden onset complaint of abdominal pain and refusal to go to school in her 17-year-old son with Down syndrome, though ordinarily he was very excited by going there [2] (pp. 167–173). This lasted for a week, until changes were made in his after-school work environment. Only much later was it learned that he probably had been sexually abused by a coworker.

Abdominal pain is often the presenting complaint of a child whose actual problem is psychological, while the complaint activates a differential diagnosis of physiological causes and may precipitate a complex fruitless workup (Chap. 5). Because of the impaired ability to articulate feelings and concepts, a person with intellectual disability may be more prone to somatization.

Case Study 1

Jane is 16 years old and affected by Down syndrome; she is customarily even-tempered, affectionate, and cooperative as a member of the family. At school, she is typically trouble-free and generally beloved by teachers and fellow students. Over the last few months, something has changed. Jane has become listless, gained weight, and has resisted invitations to do things that she usually enjoys. She resists advances from her friends at school as well as from her parents and siblings. When asked to explain her unaccustomed lack of interest in life and its activities, she offers non-specific remarks about feeling tired and disinterested. At her annual visit to her primary care physician, who is new to the family this year because their previous pediatrician retired, her mother expresses concern about these changes. Jane has no complaints, and her mother cannot offer any objective data except that Jane's menses have been less regular recently than they had been for the past 3 years since menarche.

The family is preparing for the wedding of Jane's older brother Jim, who graduated from college 2 years ago and has opened a successful small business.

Questions

1. Which of the following is the most likely explanation for this change in customary behavior patterns?
   (a) Jane has developed hypothyroidism
   (b) Jane is manifesting early dementia
   (c) Jane is simply becoming a sullen teenager
   (d) Jane is being sexually abused by someone
2. While this problem is being evaluated, Jane rather suddenly begins to complain that she does not want to go to school and that she does not like any of her classmates anymore because they are not treating her kindly. Her parents should do which of the following?
   (a) Punish Jane until she becomes more compliant
   (b) Open a discussion about her feelings regarding the fact that her classmates are all getting their drivers licenses and that this might make her feel left out
   (c) Explore the possibilities of home schooling
   (d) Ignore Jane's complaints and keep dropping her off at school each day
3. Jane has been accustomed to seeing Jim as her protector and best friend. A few days before the wedding, she becomes short-tempered and refuses to do her usual household chores. When pressed to explain her behavior, she blurts out that she hates her future sister-in-law and that she does not want to attend the wedding. Her parents should:
   (a) Drop the subject and say nothing more about it
   (b) Punish her for not doing her chores
   (c) Ask Jim to postpone the wedding until Jane can be reconciled to it
   (d) Offer to Jane the observation that everyone in the family will miss having Jim around because his presence has been such an important part of their lives

Answers

1. (a): Hypothyroidism is a common complication of Down syndrome and will often present with decreased energy, increased weight, and depression of various physiologic processes such as menses; the primary care physician should be monitoring for it as part of annual evaluations.
2. (b): Sadness and acting out may be hallmarks of depression or jealousy, which a person with intellectual disability may have difficulty in articulating. As such it is a normal part of living with intellectual disability. Ignoring it or punishing it will be counterproductive, and further isolating Jane by home schooling would certainly not be constructive. Discussion with a psychologist, pastor, social worker, or other person who can help bring sadness and a sense of disenfranchisement to the surface for discussion will be the best approach. Formal assessment and medical treatment for clinical depression may be essential in this setting.
3. (d): Honoring Jane's feelings as being normal and understandable is an important way to acknowledge her being a person whose feelings matter to her family. Jim and his fiancée are more likely to be interested in participating

Fig. 13.2 A girl with 22q deletion syndrome visiting the zoo

in that effort than they are to seriously consider postponement of their wedding. Punishing Jane or ignoring her send the signal that her feelings are unimportant; that will only worsen her isolation and pain.

22q11 Deletion Syndrome (= Velo-Cardio-Facial Syndrome, = DiGeorge Syndrome)

This condition also stems from an aneuploid state: the loss of a portion of the long arm of chromosome 22, involving up to several dozen genes that are thus rendered hemizygous (only one copy present when normally there are two copies). The cardinal features of 22q deletion syndrome are cleft palate (sometimes obvious and at other times more subtle involving only weakness of the posterior soft palate) and cardiovascular malformations (sometimes life threatening and sometimes mild). Other important common components are as far ranging as immune deficiency, learning disability, and schizophrenia (as well as a long list of dysmorphic features) (Fig. 13.2). Even among affected members of a given family there can be considerable variability such that the theoretically straightforward dominant inheritance (50 % chance of passage to each child of an affected person) may be difficult to discern among members of the extended family [3].

About 70 % of persons affected by this syndrome have some degree of speech impairment because of involvement of the soft palate (velum). Hypernasality predominates on the list of impairments. Specific strategies are in place to address and solve this problem so that affected persons need not experience social isolation, stigmatization, or career limitations as a result of this avoidable syndrome feature. Much of the second volume of Velo-Cardio-Facial Syndrome, by R. J. Shprintzen and K. J. Golding-Kushner (2011) is devoted to those therapeutic strategies [4].

Speech impairment may not be apparent for many months into infancy. A more immediate neonatal consequence of velopharyngeal insufficiency may be failure to thrive despite parents' best efforts to provide nutrition. This is a major way in which 22q11 deletion syndrome presents in the absence of obvious facial clefting. In particular, significant cardiac involvement may render an infant readily fatigued when suck and swallow require more than the usual expenditure of effort.

The speech problems inherent in 22q11 deletion syndrome constitute a functional symptom in that they create an unpleasant experience for the affected person and impair his or her function in society (because they advertise the person as "other" and contribute to isolation and discrimination by others). The problem is made more complex by the fact that persons with 22q11

deletion syndrome are also prone to intellectual delays and also to psychosis.

Case Study 2

George had considerable difficulties with infections during his early childhood. A moderate immune deficiency, associated with depressed T-cell counts, gradually resolved over his first decade. His health and development were otherwise entirely normal. In college, he married Tina (who is an advanced student of voice and plans on a career in opera). At age 21, George received a diagnosis of schizophrenia after a series of disturbing events which involved acute psychosis and hospitalization; he has been doing well with his medication regimen. Their infant son Billy, born 5 months later, had a heart murmur that has been found to be due to a small ventricular septal defect. Billy was expected to resolve this problem spontaneously over his first 2 years, but at 3 months of age he presents with "failure to thrive," having gained no weight since 1 month of age.

Questions

1. Billy's pediatrician should:
   (a) Call social services on suspicion of parental neglect
   (b) Wait another month before intervening
   (c) Send for a study of suspected 22q11.2 deletion
   (d) Send for that study but also hospitalize for intensive investigation
2. The laboratory reports 22q11.2 deletion in Billy and then documents it in George too. George's father is 82 years of age, and his mother is 55. Neither of them has ever had any health problem referable to VCFS, and there is nothing in either side of his family to suggest that the condition has been present before his birth. This means that:
   (a) George's father probably is not the source of his paternal DNA
   (b) The diagnosis is incorrect
   (c) George represents a new mutation for loss of the 22q11.2 region
   (d) In this case the clinical condition is a phenocopy (something that looks like 22q11 deletion syndrome but is actually inherited as an autosomal recessive condition)
3. When Tina and George learn about the diagnosis, they both set about reading extensively about 22q11 deletion syndrome. George experienced normal intellectual development and his schizophrenia is well controlled. Billy is now growing well after appropriate interventions to circumvent the feeding problems associated with his velopharyngyeal insufficiency, so they have been in an optimistic mood about his prognosis. Now Tina is distraught to think that her son may never share the joys of vocal music that are so important to her life. The pediatrician should:
   (a) Encourage Tina that referral to specialists experienced in the speech aspects of VCFS will probably insure that Billy has a very good outcome in speech skills
   (b) Urge Tina to abandon any hope that Billy would every sing effectively
   (c) Scold Tina for her focus on trivialities when all involved in Billy's care should be anticipating the potential onset of immune deficiency and its consequences
   (d) Distract Tina by emphasizing the dangers associated with Billy's ventricular septal defect

Answers

1. (d): There is good reason to suspect VCFS inherited from George, but this child should be afforded an immediate thorough workup for other possible causes of failure to thrive. Once the diagnosis of VCFS has been documented, velopharyngeal insufficiency with its many manifestations may be understood as a functional symptom of the underlying condition since it is not inevitable in affected persons but can be addressed with more or less success with therapeutic and learning interventions.
2. (c): While non-paternity is a possibility, new mutations are a major source of new genetic disorders of all kinds, but particularly for those which follow autosomal dominant patterns. Such events have increased risk of occurrence when paternal age is over 40, and

George's father was 60 at the time of George's birth. Neither (b) nor (d) is credible since the diagnosis has been confirmed at 22q11.2 deletion.

3. (a): When addressed by specialists with appropriate skills and experience, the problems associated with veloinsufficiency are usually quite manageable and so that is good reason to be optimistic in this matter. Neither (c) nor (d) should be actual choices for any physician.

Phenylketonuria

Inborn errors of metabolism are caused by biochemical abnormalities with more or less severe consequences to the physical and intellectual health of affected persons. Each disorder in this large family of genetic conditions is based on loss (or diminution or sometimes even gain) of function of a gene responsible for production of an enzyme, which in turn has a role to play in the construction and/or remodeling of cellular components. The most common of all these conditions is phenylketonuria (PKU). It is as prevalent as 1 in 2,600 newborn Turks, but seen in 0.01 % of newborn Irish children, and has about overall 1 in 15,000 newborn Americans from a mixture of ethnic backgrounds. PKU is inherited as an autosomal recessive trait (McKusick catalog number 261600) due to mutations in the phenylalanine hydroxylase (*PAH*) gene. Autosomal recessive traits are caused by genes located on one of the 22 numbered pairs of chromosomes, and they occur as a health problem when both parents contribute a copy of the gene which does not function normally; a person is clinically affected only when both of his/her gene copies are abnormal, but not when only one of them is (Fig. 13.1) [5].

The inability to process phenylalanine (converting it into tyrosine) leads to accumulation of byproducts that are toxic to neuron maturation. This leads to intellectual disability in almost all untreated affected persons, with intelligence scores in the range of about 50. Affected persons are typically fair skinned with blond hair. The clinical name for the condition derives from the observation of phenylketones in the urine. One of the triumphs of genetics in medicine was the discovery that a diet restricted in phenylalanine content allows a child affected by PKU to develop normally, provided the diet is instituted in the early days of life. This insight brought most of the developed world to mandatory newborn screening, so that affected infants are identified early and placed on the effective regimen that allows them a normal future. Some affected persons have mild learning disabilities even when following the dietary limitations.

Among the neurologic signs often seen in affected persons, one must take note of tremor, pruritis even in the absence of apparent dermatitis, and rhythmic rocking. These all may be thought of as "functional symptoms" because they may be disturbing to the patient and/or the patient's parents, but they are not per se debilitating or handicapping. These symptoms may become more apparent if phenylketone levels are elevated [6, 7].

Case Study 3

Dr. Freebush is consulted by Thomas, a 21-year-old man who is new to his practice. Thomas moved to town recently to work in the construction industry, which has been improving here lately. He describes himself as a migrant worker, unmarried, and "footloose." He complains of a progressively worsening tremor and pruritis of the torso.

Thomas gives a past medical history with no surgery or hospitalization and no major illnesses. He has never been prescribed any medication other than occasional antibiotics for an infection such as sinusitis. He states that he dropped out of school at the 10th grade after having had some difficulty with learning disabilities. He describes a childhood that was normal except that his parents insisted on strict dietary limitations which they told him were essential to his well-being; he abandoned those limitations when he moved away from home at 18 years of age.

He gives a family history of hypertension in his father, age 51; diabetes in his mother, age 49. His sisters, ages 24 and 16, are healthy and well and both strong students. He does not recall either of them having been required to follow his

dietary regimen. He has not stayed in touch with the family for the past 3 years.

Physical examination is notable for stable vital signs, height, and weight in the mid-normal range, fair skin with light blond hair and blue eyes. Thomas is anatomically normal. There is no sign of any dermatologic condition that might cause pruritis. The neurologic examination is normal except for a coarse resting tremor that does not interfere with movement or activity (whether gross such as lifting a chair or fine such as signing his name).

Questions

1. Which of the following is NOT a useful course of action for Dr. Freebush?
   (a) Dismiss the complaints as having no medical significance
   (b) Order complete blood count and comprehensive metabolic panel
   (c) Order plasma amino acids
   (d) Obtain medical records from his previous pediatrician
2. Which of the following is an appropriate therapeutic action?
   (a) Obtain a brain CT to investigate neurological disorder
   (b) Perform a skin biopsy to investigate pruritis without apparent dermatitis
   (c) Explain to Thomas that there is nothing to be done for his complaints
   (d) Encourage Thomas to follow a low-phenylalanine diet
3. Which of the following would not be a prudent approach to helping Thomas maintain a more healthful diet regimen?
   (a) Cognitive behavioral therapy
   (b) Self-hypnosis
   (c) Prescription of an anxiolytic
   (d) Education about PKU

Answers

1. (a): The typical intake panels (CBC and CMP) are appropriate, but the history and physical examination are consistent with PKU so that a plasma amino acid panel can be diagnostic and the pediatric medical records are likely to be consistent with this suspected diagnosis. Failing to take the complaints seriously is inappropriate.
2. (d): Brain CT and skin biopsy are invasive and expensive and will not elucidate the situation. There actually is something to be done for Thomas: low-phenylalanine diet should reduce his tremor and pruritis.
3. (c): Empowering Thomas by providing him better tools to gain control over his own behavior will be far more adaptive than depending on pharmacologic intervention that may in itself develop into a behavioral health problem over time.

Treatment

Case 1

A new physician for a child with Down syndrome may not have had the opportunity to consult the important protocol of screening recommended by the American Academy of Pediatrics [8]. She should do so and will note that hypothyroidism is a common treatable complication. Presentation with weight gain, listlessness, and loss of interest in usual activities fits the classic picture of this condition. Counseling is just as important and potentially useful to a person with intellectual disability as it is to anyone else. Resentment and sadness over inability to participate in normal milestones such as the achievement of a drivers license can be expected to have a serious impact for a teen with Down syndrome. So too can major family transitions such as the “loss” of a beloved brother (to his new wife). In both instances, consultation with a social worker, clergy person, or psychologist can be very helpful.

Case 2

The physician should explain to Tina and George that their son Billy has inherited from his father a genetic condition that is highly variable. Thus, no one involved should feel guilty for having failed to recognize the problem until the child provided

a more typical presentation than the father. The 50 % risk for transmission to any new child should be made clear, and an empiric approach to treatment of signs and symptoms promised. Tina should be assured that, while perfect diction might not be possible, there is no need to despair of Billy having a pleasant speaking and singing voice if the appropriate specialists are consulted to help him avoid developing bad habits and promote good diction.

Case 3

Thomas should be gently but clearly instructed on the importance of limiting phenylalanine ingestion. Consultation with a geneticist and specialized nutritionist will allow him to identify ways in which he can modify his diet without necessarily limiting it so strictly as to bring back unpleasant memories of his parents' restrictions (which must be much more strict during childhood than later). Dr. Freebush might encourage a more disciplined approach to diet by pointing out that tremor can make a person appear less than desirable to hire for a highly physical occupation such as carpentry. Thus the increased discipline can be expected to help him stay gainfully employed.

Conclusion

Genetic disorders involve every part of the body, at every stage of development from conception to the grave. Functional symptoms in genetic disease can be found across a broad spectrum. They are not really the functional symptoms of genetic disease but the functional symptoms of a specific disease that happens to be genetic. Intellectual disability is listed as a component of over 2,000 syndromes with genetic underpinnings [9]. The specific components of intellectual disability are highly varied and complex (including attention-deficit disorder, autism spectrum, self-injurious behavior). They also overlap in complex ways with mood disorders, thought disorders, aggression, etc. Each of these must be addressed according to its exact presentation, with treatment that addresses its actual characteristics rather than any hypothetical or genetic framework. The functional symptoms of genetic disorders that affect cardiac, renal, pulmonary, skeletal, endocrine, or neurologic functions are also highly varied. The physician confronted by a patient with complex signs and symptoms due to underlying genetic disease may find it challenging to recognize each for its actual implications. It is important not to respond disproportionately to it on the basis of the patient's discomfort, the parents' worry, or the clinician's unfamiliarity with the diagnosis.

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Functional Symptoms in Children Who Have Developmental or Behavioral Differences

14

Paul G. Taylor

Abstract

Children and adolescents who have developmental or behavioral differences (DBD) may present with the same range of nonorganic functional symptoms as otherwise typical young people. Paradoxically, DBD may itself be a nonorganic functional symptom. The presence of DBD may introduce potential pitfalls in the assessment and management of functional symptoms, leading to incomplete or erroneous conclusions and inappropriate interventions. Some specific nonorganic functional symptoms are more likely to present in children with developmental differences (DD) compared to typically developing children. Children who have behavioral differences (BD), particularly autism spectrum disorder, are more likely to present with functional symptoms than their neurotypical peers. Failure to take DBD into account often leads to hasty and incomplete evaluation of symptoms and consequent failure to address key underlying problems. An approach to diagnosis and treatment which takes DBD into account is likely to lead to the best outcome for the affected child and family. Included in this discussion are several symptoms which are, strictly speaking, not functional symptoms but which occur more often in association with DBD than in typical people. These symptoms may be ameliorated with effective management of the underlying DBD.

Keywords

Developmental difference • Behavioral difference • Autism spectrum disorder • Anxiety • Paroxysmal non-epileptic phenomena • Selective mutism • Alopecia areata • Trichotillomania • Attention-deficit hyperactivity disorder • Tourette syndrome

Introduction

First and foremost it is useful to bear in mind that children and adolescents with DBD may manifest many of the symptoms covered elsewhere

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in this book. For example, a 9-year-old with a significant developmental difference who presents with a clear history of recurrent abdominal pain which conforms to the Rome II criteria for irritable bowel syndrome (Chap. 5) may be assessed and managed according to accepted guidelines. The same is true of other discrete presentations.

However, there is good evidence that the presence of DBD predisposes to functional symptomatology which reflects factors unique to this group of patients. This knowledge can be helpful in clarifying and refining the diagnosis of functional disease in a patient with DBD. Furthermore, a presenting functional symptom may itself be the sole symptom which points to an underlying DBD, which the astute clinician may go on to confirm. For example, the presence of repetitive behavior or of generalized anxiety may lead to the diagnosis of autism spectrum disorder (ASD) and, in turn, to specific opportunities for wider effective interventions, which not only deal with the presenting symptom but also lead to amelioration in many other aspects of the life of the person with a DBD.

Functional Symptoms as the Expression of an Impoverished Environment

Any impoverished environment is complex. Impoverishment can include lack of physical resources necessary for survival including shelter, food and clothing, and lack of social and emotional resources necessary for optimal personal development. The victims of impoverishment are adults and children. The effects of impoverishment on adults may diminish the capacity of the adult to provide the social and emotional support necessary for the child's best development. Children reared in an impoverished environment may present with either or both of two dramatic functional symptoms: growth failure and intellectual impairment, which often coexist.

Nonorganic Growth Failure Due to an Impoverished Environment

The existence of growth failure due to emotional and environmental deprivation is well documented [1]. Studies have shown that environmental impoverishment may be the leading cause of nonorganic growth failure throughout the world. In developed countries, the incidence of nonorganic growth failure in children seen in primary care settings is reported to be 5–10 % [2] and 1–2 % of all admissions to hospital [3]. Nonorganic failure to thrive is diagnosed in at least 75 % of all children admitted for assessment of growth failure.

Growth failure may occur because of nutritional deficiency associated with impoverishment. Lack of maternal attachment leading to emotional deprivation also leads to growth failure [4]. There are many reasons why this may occur including the presence of maternal psychological health issues, learning disability, substance abuse, and other health concerns.

On the other hand, the behavior of the infant him or herself may lead to failure to thrive. A common situation that illustrates this is the otherwise well term breast-fed infant who becomes progressively more lethargic and less demanding for feeds. With appropriate intervention this downward spiral can be quickly reversed. Persistent refusal to suck, failure to indicate hunger, and refusal to eat presented food may occur. Some infants may experience distressing escape of fluid from the pharynx into the larynx during swallowing leading to refusal to suck. Evaluation of the antecedents of failure to thrive associated with an impoverished environment, therefore, mandates assessment of the role of the infant's behavior as well as that of the mother [5].

The diagnosis of growth failure depends on interpretation of available anthropomorphic data. There is no single universally accepted definition of growth failure. A weight to length or height ratio <70 % of the predicted value may be the most useful single indicator, but review of multiple growth data is necessary to inform a diagnosis.

A diagnosis of nonorganic growth failure is usually self-evident after the obtaining of a detailed history, with special attention to comprehensive assessment of the child's environment and feeding patterns, a careful physical examination to exclude signs of organic disease, and interpretation of growth data. Unless otherwise indicated, batteries of laboratory tests and imaging procedures are not necessary to make a diagnosis; that is, nonorganic failure to thrive need not be a diagnosis of exclusion.

Admission to hospital is usually recommended in order to observe the child's feeding patterns, allow free access to nourishment, and to permit a more detailed assessment of the physical, social, and psychological elements that are present in the child's home environment. Central to this is a careful assessment of the mother's physical, emotional, and behavioral health. This process is usually rewarded by rapid weight gain in the infant and rehabilitation of the maternal–infant relationship. However, although such a single intervention may reverse the process, the risk of relapse is high. This mandates ongoing surveillance of the child and family and steps to rescue the child on a temporary or permanent basis if necessary.

Developmental Disruption Due to an Impoverished Environment

Numerous sources confirm that an impoverished environment can interrupt normal development of infants and children [6]. Such impoverishment may be primarily economic [7] or psychological [8], although both are likely to be present together [9].

The term "reactive attachment disorder," a diagnostic entity described in the Diagnostic and Statistical Manual of Mental Disorders third edition (DSM-3) [10], the Diagnostic and Statistical Manual of Mental Disorders fourth revised edition (DSM4-R), and International Classification of Diseases, tenth edition (ICD-10), may be used to describe this constellation of abnormal reactions to an impoverished environment [11, 12]. The fifth edition of the DSM published in 2013 maintains this diagnostic framework and proposes specific criteria which describe the pattern of disrupted development and the predisposing environmental disturbances on which a diagnosis can be made. In addition, the DSM-5 delineates a second discrete pattern of abnormal development occurring in response to environmental impoverishment in which disinhibited social engagement occurs.

The effects of environmental impoverishment may be ameliorated by returning the child to a consistent and nurturing environment. However, studies of the long-term outcome of such children show that cognitive and emotional deficits may persist. In a follow up of 111 children adopted into UK homes from Rumanian orphanages, 6 % of children showed enduring autistic-like behaviors compared to none in 51 infants born in UK and adopted within the first 6 months of life [13]. In a follow-up study of cognitive outcomes at age 11, of 131 Rumanian children adopted from institutions, compared to 50 UK adopted children, marked adverse effects were found in many of the children who were over 6 months of age on arrival in the UK. Of these children, those with the highest degree of impairment on arrival did display some catch-up of development. Nevertheless, a deficit of 15 IQ points persisted in the whole group compared to the UK adoptees [14]. These findings contrast with follow-up studies of infants presenting with failure to thrive without environmental deprivation. In these studies minor but persisting growth deficits were documented in some children associated with a collective cognitive deficit averaging only three IQ points compared to controls, a difference of dubious practical significance [15].

In a study of 10 Rumanian orphans, mean age 8.8 years, who showed mild neurocognitive impairment, impulsivity, and attentional and social deficits, positron emission tomography showed significantly bilateral decreased glucose metabolism in the orbital frontal gyrus, the infralimbic prefrontal cortex, the medial temporal structures (amygdala and head of hippocampus),

the lateral temporal cortex, and the brain stem compared the normal cerebral hemisphere in a control group who were investigated for focal epilepsy [16]. The same investigators went on to report abnormal brain connectivity in a small group of survivors of eastern European orphanages using diffusion tensor imaging, which uses a magnetic resonance imaging technique to display neural tracts. Compared to a control group, structural changes in the left uncinate fasciculus were documented in children who had experienced socioemotional deprivation. The investigators postulate that these changes may, in part, underlie the persistent cognitive, socioemotional, and behavioral difficulties that are commonly observed in these children [17].

Functional Symptoms That Have Been Shown to Occur More Frequently with Developmental Delay

Paroxysmal Non-epileptic Phenomena

Evidence suggests that pseudoseizures, more appropriately termed paroxysmal non-epileptiform phenomena (PNEP), occur more frequently in children and adolescents who have concomitant developmental delay or impaired cognition [18]. Furthermore, PNEP may occur more frequently in young people who have a history of childhood abuse, disturbed family background or family history of psychiatric disorder. PNEP is more common in females than males and often occurs in young people who also have a seizure disorder confirmed by electroencephalogram or in association with a history of head injury [19, 20]. The diagnosis of PNEP is usually clear from the history (see Chap. 2) and does not usually require additional investigation.

The long-term outlook for children who present with PNEP is encouraging. Of 35 children who attended a tertiary pediatric neurology center 66 % became PNEP free. In a further 23 % a reduction in frequency of PNEP of over 50 % was recorded. In only two of the study children did PNEP persist unchanged. The outlook was best for children whose PNEP was not accompanied by epilepsy [21].

Functional Symptoms Associated with Behavioral Differences

Anxiety and Depression

There is extensive evidence linking psychiatric symptoms, especially anxiety and depression with functional symptoms in childhood and adolescence, such as abdominal pain and headache. Long-term studies indicate that functional symptoms in childhood may predict psychiatric illness later in life. The personality profiles of children with functional symptoms include perfectionistic and obsessive traits, undue sensitivity and feelings of insecurity, and excessive worrying. These traits may predispose the young person to anxiety disorder. Children with functional symptoms show relative paucity of effective coping skills compared to children who have the same symptoms due to organic disease and to children without symptoms [22].

The level of distress shown by children with functional symptoms can be increased when parents respond to the symptoms in a concerned and overly sympathetic manner that provides unintentional reinforcement of the symptoms. This creates the potential to intervene by counseling parents to take a more pragmatic approach in their response [23].

Functional Symptoms in Autism Spectrum Disorder

ASD is common. In a recent large population-based study, the incidence of ASD was found to be just below 3 % of all children [24]. The diagnosis was previously not recognized in two-third of school-aged children who had autism and normal academic intelligence [24]. The distribution of academic intelligence in ASD follows a bell-shaped curve. Autism associated with exceptional intelligence is often loosely referred to as

Asperger syndrome. When it is associated with significant impairment of cognitive ability this creates the clinical picture of classical autism, which is how many of the general population, including health professionals and teachers, think about autism. This hampers the effective recognition and management of infants, children, and adolescents who have autism and normal intelligence [25].

General anxiety and situation-specific anxiety complicate the lives of children, adolescents, and adults who have ASD as defined according to DSM-5 criteria. This has been well studied and described in the literature [25]. In one study children with ASD aged 3–5 years who had anxiety ($n = 182$) outnumbered those who did not (135). At age 6–12 years the ratio was 301:191. Children with autism and high intelligence were more likely to be anxious than those with average or low intelligence. Anxiety was more prevalent at school than at home [26]. In another study of children aged 10–12 years who had ASD, at least 70 % had additional diagnoses, including social anxiety in 30 % of the cohort [27]. In a novel reverse approach, Towbin and colleagues studied a group of 93 children, mean age 12.7 years, referred for assessment of symptoms of anxiety. ASD was diagnosed for the first time in 62 % of this cohort [28].

Anxiety associated with ASD often leads to behavior mistakenly labeled as anger or aggression. This is particularly true of children with autism and normal academic intelligence attending grade school. The presence of normal academic intelligence blinds many, if not most, school professionals to the possibility of autism in school age children. As a consequence, children with unrecognized autism are subject to conventional interpretation of their behavior and interventions designed for children who do not have autism. Neither of these is helpful or effective when autism underlies behavior [25]. When autism is recognized, the most appropriate response is a thorough assessment of the child's environment at home and at school in order to identify those aspects of the environment that cause anxiety and which can be easily modified to accommodate the person with autism. Most importantly, these include the knowledge, attitudes, and behaviors of the adults in the child's environment. This is much more effective than trying to change the child by employing conventional behavioral management techniques.

Treatments proposed for anxiety states in children and adults include cognitive behavioral therapy (CBT), anxiolytic medication, hypnotherapy, and a variety of alternative and complementary approaches. Very little of the research done in this area is specific for people with autism, especially children. In one of only two studies published on the effectiveness of CBT in children with autism, Wood and colleagues showed that of 36 of 47 children with high-functioning autism who completed 16 multimodal CBT sessions 78 % showed improvement compared to 8.7 % of children assigned to a waiting list [29]. In 71 children with Asperger syndrome aged 10–12 years who were randomly assigned to receive CBT alone, CBT with their parents, or no intervention, positive changes were more frequent in those who received CBT with their parents, compared to those who received CBT alone. Both groups did better than the group assigned no treatment [30].

Studies regarding the use of anxiolytic medication for children, young people, and adults with ASD highlight the modest gains and high incidence of side effects [31–33]. In contrast, in a study of 17 children and adults who had significant anxiety associated with autism, all responded to a single intervention using clinical hypnosis to target specific situational anxiety or generalized anxiety. Improvement was maintained for up to 17 months follow-up in all subjects [34].

ASD can also be related to failure to thrive. In a small cohort of six children with feeding difficulties leading to failure to thrive, the underlying diagnosis was found to be autism in all of the cases [35].

Selective Mutism

Selective mutism has not been well studied. The incidence of selective mutism is reported to be 0.9 % in one study [36]. As early as 1989 the association between autism and selective mutism

was identified [37]. Subsequent reports have not specifically examined this association. However, a more recent study of selective mutism in 54 children compared to a control group of 108 normal children showed that of the children with selective mutism 68.5 % met the criteria for a diagnosis reflecting developmental disorder/delay compared with 13.0 % in the control group. The criteria for any anxiety diagnosis were met by 74.1 % in the study group versus 7.4 % in the control group. In the study group 46.3 % of the children met the criteria for both an anxiety diagnosis and a diagnosis reflecting developmental disorder/delay versus 0.9 % in the controls [38].

By inference from the study of Towbin and colleagues cited above, those children with selective mutism occurring in association with anxiety may have an incidence of unrecognized ASD of up to 62 % [28].

Alopecia Areata

Alopecia areata is an enigmatic phenomenon of unknown cause. It is postulated that altered T-cell auto-immunostasis is involved in the etiology. Studies of alopecia areata document the high incidence of anxiety in people with this condition. In one study of 80 patients, including young people, with alopecia areata lasting an average of 7.8 years, more than three-quarters of patients believed that stress was the cause of their hair loss [39]. Alopecia areata is uncommon in children and is most often the result of trichotillomania. Numerous treatments are proferred for alopecia areata. Most are based on interventions designed to alter the host immune response, such as topical, locally injected, or systemic steroids. However, a recent Cochrane review was unable to identify any robust scientific evidence to support the efficacy of any medical interventions [40]. Numerous complementary and alternative interventions are also proposed to cure alopecia areata. Again there is no evidence of efficacy for any of these proposed interventions [41].

Trichotillomania

Trichotillomania, or hair pulling, is a common functional symptom seen in children, adolescents, and adults [42, 43]. In many cases this can lead to alopecia areata and trichobezoar when the dislodged hair is ingested. Trichotillomania is frequently associated with internalizing symptoms and psychiatric diagnoses, most commonly obsessive compulsive disorder and chronic anxiety, although these disorders are not universally present [44]. In a cohort of adult females with trichotillomania, the incidence of childhood trauma in general and emotional neglect in particular was significantly greater compared to a control group [45].

Treatment for trichotillomania generally involves some sort of behavioral therapy. This produces at least short-term improvement in symptoms across all age groups, including children as young as 7 years [46]. Clinical hypnosis (Chap. 21) also is an option for treatment.

Functional Symptoms in Attention-Deficit Hyperactivity Disorder

Attention-deficit hyperactivity disorder (ADHD) may be complicated by functional symptoms in at least 60 % of cases. These complicating symptoms are usually behavioral in nature and may be regarded as functional in the sense that there is no obvious organic basis for any of them. Functional behavioral complications of ADHD include oppositional-defiant disorder, conduct disorder, anxiety disorder, and depression. In a small proportion of children who present with symptoms of ADHD, the underlying diagnosis will turn out to be childhood bipolar disorder, although this is not common and remains a disputed diagnosis. Further discussion of these functional symptoms associated with ADHD is outside the scope of this text.

ADHD and autism coexist more frequently than would be expected from their individual incidence [47]. In patients diagnosed with inattentive subtype of ADHD, assessment should be

extended to exclude the possibility of autism associated with normal intelligence. This will turn out to be the correct diagnosis most of the time (personal observation).

Tourette Syndrome and Attention-Deficit Hyperactivity Disorder

Tourette syndrome, a tic disorder characterized by involuntary physical movements and vocal utterances, is known to occur more frequently in association with ADHD and with autism than in the general population. This can be a disruptive association. An instructive study looked at 207 children aged between 7 and 18 years, including 144 boys and 63 girls. Forty-two of these children had uncomplicated Tourette syndrome and 52 had uncomplicated ADHD. Fifty-two children had both ADHD and Tourette syndrome. A control group of 61 children had neither diagnosis. Children with only Tourette syndrome showed the same low profile of aggressive or delinquent behavior as the control group. Children in whom Tourette syndrome was complicated by ADHD displayed the same level of disruptive behavior as children with ADHD alone. However, taken in entirety, the presence of ADHD in children with Tourette syndrome added significant further burden on the families of affected children [48].

Case 1

Evangelina (not the patient's real name) was first seen at the age of 11 because of a short history of running away from home without an obvious reason. She is the only daughter of a separated mother who works in a supermarket. Evangelina was said to have learning difficulties although a formal cognitive assessment had never been undertaken. Review of previous medical history revealed the presence of generalized tonic–clonic seizures well controlled on anticonvulsant medication and difficulty in forming and maintaining friendships with children her own age. Clinical examination revealed a child with a somewhat blank and expressionless facial response to interaction with no further abnormal physical signs. Focused questioning revealed a history of dramatic tantrums in earlier childhood, difficulty paying attention, a predilection for daydreaming, difficulty following complex instructions, and a lack of two-way emotional interaction with her mother. Following further formal behavioral assessment, a diagnosis of ASD associated with chronic anxiety and low average intelligence was added to that of her seizure disorder.

Appropriate counseling on how to create a home and school environment supportive of children with ASD was given to Evangelina's mother and to her school. This resulted in improved adaptive behaviors.

As Evangelina progressed through high school tonic clonic seizures recurred. These always occurred at school and resulted in urgent conveyance by ambulance to the local hospital emergency department. Seizure activity had always ceased prior to arrival at the emergency department. Review of Evangelina's circumstances showed that she had become a member of a small group of girls at school but was often shunned by them. Her peer group, however, were solicitous of her seizures and would always congregate to assist during a seizure.

Her mother and pediatrician became disillusioned with frequent calls from school to attend to Evangelina at the emergency department. Neither believed that these seizures warranted such an extreme response. At a school meeting of those professionals involved with Evangelina, including the pediatrician, chaired by the school's special needs coordinator, it was agreed that whenever a seizure occurred in future her group of friends would walk away and the school would not respond by calling an ambulance. Evangelina would be left to recover by herself in the school's infirmary.

Within 3 months Evangelina became seizure free and has remained so for the past 3 years. She does, however, continue to take her regular anticonvulsant medication. She is now working as a supermarket check-out clerk.

Questions

1. Factors in the history of this girl consistent with a diagnosis of paroxysmal non-epileptic phenomena include (more than one may be correct):
   (a) Previous history of EEG confirmed tonic–clonic epilepsy
   (b) Absence of seizures outside school
   (c) The presence of a degree of mild intellectual impairment
   (d) Coexisting ASD
   (e) All of the above
2. Which of the following statements regarding paroxysmal non-epileptic phenomena is true?
   (a) More than 50 % of cases of PNEP undergo spontaneous resolution
   (b) PNEP is commonly associated with recurrent abdominal pain
   (c) PNEP is more common in boys than girls
   (d) PNEP is associated with recall of the event
   (e) All of the above
3. The assessment of PNEP must include which of the following?
   (a) Video-electroencephalography
   (b) Overnight sleep study
   (c) Assessment of calcium metabolism
   (d) Evaluation of the hypothalamic–pituitary–adrenal axis.
   (e) None of the above

Answers

1. (a, b, and c): In this clinical vignette Evangelina displays the symptoms of a paroxysmal non-epileptiform phenomenon (PNEP). The relevant factors in her history include the fact that these events occur only at school in a person with a mild intellectual disability and a previous history of EEG confirmed tonic–clonic epilepsy. However the history of EEG confirmed seizure disorder does confound the situation by leading others to conclude that the events in question represent break through epileptic seizures requiring urgent medical intervention. The presence of ASD is irrelevant.
2. (a): It is encouraging to note that spontaneous resolution is the usual outcome of PNEP, which occurs more commonly in girls than boys. There is no recognized relationship between PNEP and recurrent abdominal pain, nor any recall of the event.
3. (e): The diagnosis of PNEP can be confidently made on clinical grounds alone. Ancillary investigations are not necessary and only serve to increase the cost of good care.

Case 2

Milton (not his real name), aged 9 years, was brought to a regional center because of his behavior at school. He seemed to be constantly in trouble. He would be found fighting other children. Milton would often lash out for no particular reason. This seemed to be indiscriminate, without provocation and often directed toward the nearest person, which could be a boy or a girl. Milton had been expelled from two previous schools and was facing the same sanction at his current school.

Further history revealed that Milton lived with his mother but would spend time with his father, a man of strict religious beliefs who was intolerant of misbehavior. He had more or less disowned Milton. Milton had no friends. He had never engaged in imaginative play with other children but would spend hours playing alone with imaginary people who spoke a different language. When in trouble Milton would enrage the adults in attendance by smiling at them in a way which was interpreted as insolence. This tended to erode goodwill. Further history was unremarkable apart from behavioral features indicating the presence of ASD in a boy of normal intelligence.

An approach was made to his school with the challenge of making the school autism friendly. It was proposed that Milton's behavior was the result of undifferentiated anxiety secondary to his autism leading to panic reaction expressed as physical violence. An analogy was made with a cornered cat whose response to fear is to come out hissing, spitting, and slashing with exposed claws.

It was suggested that Milton's behavior would improve if he were able to reduce his feelings of anxiety. A classroom behavior management

system was put in place in which Milton was free to enter an empty fridge box placed in the corner that contained some cushions and soft material, whenever he felt uncomfortable. He could return to the class whenever he felt ready. In other words, Milton was given the freedom to manage his own feelings.

One can imagine that this was not an easy proposition to sell to the school hierarchy. They argued that there should be only one set of rules which should apply to all children. If some children were granted exceptions, one could expect all children to want the same privileges. The counter-argument was that children are perfectly capable of understanding why some of them might benefit from different rules. After all, they have known from early childhood experiences that some children are different and this never bothered them. Their natural acceptance was reinforced by the permissive attitude of early childhood educators who are skilled in inclusive strategies.

If Milton's classmates were given an explanation for why different rules apply to him, they would readily accept this and their knowledge of his differences would be affirmed. If Milton were no longer marginalized by how his behavior was managed, one could expect congruence and synergy to emerge in both teachers and children which would be supportive of Milton and assist in his adaptation.

Which is what happened. To nearly everyone's surprise and delight Milton soon stopped lashing out and was able to go from day to day without problems. He stayed at the same school and then progressed to high school along with his age peers.

This case illustrates that the most effective way to manage behavior due to autism is to manage the child's environment, not the child [24].

Questions

1. The most effective response to a child who displays random aggressive behavior is to
   (a) Discuss the rules of etiquette with the child
   (b) Invoke a sanction such as loss of privileges
   (c) Refer the child for anger management
   (d) Refer the child for a comprehensive behavioral assessment
2. All of the following statements regarding childhood autism associated with normal intelligence are true EXCEPT
   (a) The children who grow up alongside the child with autism already know there is something different about the child in question
   (b) In responding to the child with autism children take their lead from how the adults in the child's environment respond to the child
   (c) Many children who have autism associated with normal intelligence progress to adulthood without a diagnosis ever being made
   (d) Children with autism associated with normal intelligence respond well to conventional behavior management strategies
3. Functional symptoms that occur in children with autism may include which of the following (more than one may be correct)?
   (a) Headache
   (b) Abdominal pain
   (c) Lashing out
   (d) Failure to thrive
   (e) Spectacular meltdowns followed by no recall

Answers

1. (d): Random and apparently unprovoked lashing out is an unusual behavior. It requires a different response to similar behavior that is provoked or premeditated. It can be difficult to make this distinction without careful analysis of the behavior and its context. Therefore the best response to this kind of behavior is a comprehensive behavioral assessment undertaken by a professional who is trained and skilled in dealing with children who may have ASD. Answers a, b, and c. are inappropriate and would be ineffective in this situation.
2. (d): Children who grow up alongside a child with autism are aware that there are differences between themselves and the child in question but this usually does not bother them

unless the adults in their environment show intolerance of the autistic child's behavior. Recent evidence suggests that over two-thirds of school children diagnosed with autism as the result of population screening had not been previously recognized as having autism. Standard behavior management techniques employed with children are designed for children who do not have autism and rely on the presence of normal social and communication competence. The differences in social and communication behavior in children with autism make standard behavioral approaches useless. Attempts to change behavior in children with autism require an approach which accommodates the realities of autistic thinking.

3. All the options in question 3 are correct.

Case 3

Jane (not her real name), aged 12, was referred for a second opinion. She was being seen at the local child and adolescent mental health service because she refused to speak in most situations and specifically at school. The therapeutic response to this was to engage Jane's parents in a parent-training course and to construct a system of sanctions and rewards which were dependent on the amount of speech Jane produced.

Jane had also developed alopecia areata of large parts of her scalp.

On assessment Jane came with both her parents, each of whom was unenthusiastic about the treatment approach being taken by the mental health team. They described a child of above average intelligence with a passion for art and for animals. They provided a safe and nurturing home environment. They reported that Jane spoke freely at home if she had something to say. The remainder of the history was noncontributory.

On examination, Jane was unwilling to engage or be examined. She wore a headscarf that covered her entire scalp. She stared with wide eyes at the pediatrician. She indicated her response to questions by nodding or shaking her head. She sat immobile for the duration of the encounter.

Jane's parents and her school completed questionnaires designed to elicit evidence of behavioral disorders, in particular ASD. From these it was clear that the diagnosis underlying Jane's presentation was ASD and chronic anxiety.

A plan was made. Jane's involvement with the mental health team was discontinued. Jane was withdrawn from school and enrolled in a home-based school program supported and directly supervised by the regional educational authorities. She was assigned a part-time teacher who visited her at home on 2 days each week. Her parents reported that for the first time Jane appeared to them to be happy and relaxed. Her teacher reported excellent academic progress. Jane began drawing caricatures of animals, which displayed impressive artistic competence. At each visit to the pediatrician she presented him with a drawing she had made. She indicated that she now felt happy.

After 2 years Jane agreed to a plan to attend her local high school for one afternoon a week to work in the art department during a time when the art room was otherwise vacant. She would arrive while the rest of the school was in class and leave at the end of the day after the rest of the students had departed.

Jane's artwork continued to improve to the point where she began making true-life portraits of dogs from photographs sent to her by proud owners. She presented her pediatrician with one of these. This could be the basis of a future internet-based source of livelihood. By this time Jane's alopecia had fully resolved although she still preferred to wear a headscarf when not at home.

Questions

1. Which of the following statements about alopecia areata is correct?
   (a) Alopecia areata is a functional symptom associated with a history of childhood abuse and neglect
   (b) Alopecia areata is caused by refractory fungal infection of hair follicles
   (c) Alopecia areata responds well to combination topical steroid and fungicide treatment

(d) Alopecia areata remits spontaneously in all cases within 5 years of onset

2. Effective management of selective mutism includes which of the following
   (a) Habit reversal therapy
   (b) Second-generation antipsychotic medications
   (c) Consistent sanctions when there is no verbal response to questions
   (d) Comprehensive assessment of underlying psychiatric or behavioral antecedents
3. In younger people with selective mutism, which of the following approaches is appropriate
   (a) Insistence on maintaining normal day to day activities and expectations
   (b) Referral to a residential treatment program
   (c) Acceptance that departure from conventional expectations and requirements may be justified to reduce extraneous sources of anxiety
   (d) Referral for speech-language or music therapy

Answers and Discussion

1. (a): Alopecia areata is an entity of obscure etiology. There is no evidence that fungal infection underlies this condition. Therefore treatment with antifungal agents is not indicated. There is no robust evidence that treatment with steroids is efficacious. Alopecia areata can be a longstanding chronic condition. There is a recognized association between alopecia areata and childhood abuse and neglect although this is not a constant relationship.
2. (d): Selective mutism is a complex condition which requires comprehensive assessment in order to design the most useful intervention. The use of cognitive behavioral therapies or medication may be indicated in selected cases but when used routinely are more likely to worsen the symptom. The most useful intervention is often a modification of the lifestyle of the person with mutism to achieve an optimal level of comfort. Over time it is likely that the person with mutism will become more resilient with resolution of symptoms in many if not all settings.
3. (c): For the reasons outlined in the discussion of question 2.

Conclusions

Functional symptoms associated with disorders of development and behavior occur frequently. The significance of these symptoms may be underestimated. Careful analysis of how and why a child with such symptoms presents to the clinician can lead to significant improvement in the child's symptoms and the wider environment. The most effective interventions are those designed to modify the environment in which the child and family function.

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Functional Symptoms in Pediatric Emergency Medicine

15

Asalim A. Thabet

Abstract

Common disorders that frequently present as a result of functional issues in the pediatric emergency department include chronic non-pathological pain, acute chest pain, syncope, recurrent abdominal pain, and fatigue. These complaints are often broad in nature and nonspecific, leading to potentially lengthy diagnostic evaluations that are cumbersome for both the patient and the healthcare system. This chapter describes three cases of patients presenting to the emergency department whose optimal treatment required addressing their underlying psychological issues. Incorporation of child life specialists and others trained in non-pharmacologic stress reduction can assist in alleviating the anxiety and pain associated with functional issues in such patients.

Keywords

Emergency medicine • Chest pain • Abdominal pain • Syncope • Fatigue • Procedures • Psychogenic • Non-pharmacologic • Paresthesia • Postconcussion

The pediatric emergency department (ED) is the central clearinghouse for a wide variety of different medical concerns. Pediatric EDs handle everything from the most intimidating and life threatening of situations to benign self-limited processes. The main purpose of all ED providers is to rapidly identify and correct any potentially life threatening situation. Sometimes, however, a concern that appears to be life threatening and causes patients to be fearful is in fact the result of physical symptoms with a functional basis. Commonly recognized functional disorders in the emergency department can result in chronic non-pathological pain, acute chest pain, syncope, recurrent abdominal pain, and fatigue. At other times, a functional complaint may be a reflection of parental issues. For example, a divorced parent may focus on a child's symptoms as a way of proving that the other parent provides inadequate care.

Functional somatic symptoms may make an ED evaluation very difficult because of their nonspecific nature, which usually cannot be explained

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by a single disease process. As symptoms become less distinct, extensive investigations may often be undertaken. The persistence of functional symptoms can also lead to increased cost, excessive use of resources, and disability for the patient (and family). For example, costs related to the management of painful medical presentations in the USA are growing, including those associated with disability days and lost wages [1]. In addition, patients with functional symptoms who are not diagnosed or addressed properly frequently seek recurrent medical care for the same complaints. This has been associated with significantly higher rates of overall psychosocial impairment [2]. Non-pharmacological adjunctive therapies, targeted at the psychological aspects of pain, can allow for substantial cost savings [3]. Incorporation of child life specialists and others trained in non-pharmacologic stress reduction into the management offered at a pediatric ED can assist in alleviating the patients' anxiety and perceived pain [4]. Oftentimes, the best management option is a combination of both pharmacological and non-pharmacological measures to address pain and reduce overall healthcare costs.

Several factors associated with provision of health care in the ED lead to the necessity for more extensive diagnostic testing in patients with a strong likelihood of having functional disorders, as compared to evaluation of such patients in the primary care setting:

1. Patients evaluated in the ED are more likely to have serious diagnoses because of the emergent nature of their complaints.
2. In the ED, the clinician generally has no previous relationship with the patient or family. Therefore, the clinician cannot easily compare the patients' presenting complaints with their previous symptoms and physical examination findings.
3. Given the absence of a long-term relationship, families are less apt to be receptive to reassurance provided by the ED clinician.
4. As follow-up after an ED visit may not be reliable, ED evaluations need to be more comprehensive in order to exclude serious diagnoses that could be identified through regular provision of primary care.

Patients with functional symptoms who present to the ED should be discharged home with a recommendation for close follow-up with a primary provider for long-term therapies. If an individual does not have a primary provider, a primary provider should be established immediately to ensure adequate long-term resources. Without a medical home, such patients remain at high risk for frequently revisiting the ED, which leads to increased stress for patients and their families and inefficient utilization of healthcare resources.

Chronic Non-pathological Pain

Parents may feel a sense of helplessness when the child is overcome with pain that is not easily resolved with home therapies. The prevalence of recurrent pain in children is more than 30 %, and up to 50 % of children endure clinically significant amounts of pain [5–7]. Chronic pain is affected by multiple issues including biological processes, psychological factors, and sociocultural factors. The evaluation of pain is hampered when a child cannot express him/herself well due to either age or developmental delays [7].

Pain may be a result of actual, potential, or perceived pathology. Psychological and physiological assessments are necessary to determine appropriate pain management since pain represents an intricate balance between physiology and emotion [5]. Thus, pain often is the result of physical abnormalities with a functional overlay. Development of a treatment plan for the child's pain may initially require an extensive diagnostic workup. Such investigations may induce more anxiety and stress for the patient and family.

The identification of the source of pain and implementation of its treatment is essential to the empathetic care of children. Many conditions can be associated with persistent or elevated perceptions of pain. If a child has a history of a recent health condition that apparently triggered the pain, then the pain could be related to the physical result of the original health condition and/or an emotional reaction to the health condition. For example, patients in a motor vehicle collision

may develop pain resulting from the accident, but assuming no serious injury has occurred, this should dissipate within days. Assuming a normal physical exam and radiographic findings, pain related to this collision months later likely is due to functional preservation without continued pain inducing physical pathology.

Management and Treatment

Assessment of pain and its management is an important facet of pediatric emergency medical care. This assessment may be completed through self-reports and by behavioral scales that indicate levels of pain experienced. Oftentimes the medical team can provide relief of pain with objective findings, but its subjective elements may not be relieved with pain medications. In this case, it is imperative that physicians consider alternative therapies as adjuncts to standard medical care in order to alleviate subjective pain [8]. When medical personnel are unable to effectively control pain, there is an increased probability that patients will necessitate extensive and invasive investigations because the index of suspicion for a serious pathology increases.

Adequate pain control can be difficult as it oftentimes requires non-oral medication administration. This can provide additional stress, fear, and anxiety in a child who may already be sensitized to the frightening hospital environment. In some circumstances, intravenous (IV) access will need to be obtained for both medication administration and diagnostic evaluation. Peripheral IV lines are one of the leading causes of pain for children presenting to the ED [6]. Peripheral venous access in children is difficult because of their anatomy (smaller, deeper veins) and their fear of the unknown and pain related to the procedure. Children will often physically resist these interventions as a result of their fears. Peripheral IV insertion is only successful on the first attempt 53 % of the time. This number increases to just about 90 % after four attempts [9]. Repeated placement attempts lead to an increased use of resources, cost, and most importantly parental and patient distress. In addition to the psychological strain on the staff, patient, and family, failed IV placement results in delays in treatment [10]. Addressing anxiety through non-pharmacological means can result in an increased "sense" of anesthesia and ultimately increased chance of successful procedural placement [8].

Pain control for children is imperative for physiological, psychological, and emotional improvement. Uncontrolled pain can result in both long-term consequences (fear of medical facilities and heightened sensitivity to subsequent care) and short-term consequences (extended length of stay, slower healing, emotional trauma, and suffering) [11]. Pain affects each individual differently, so it must be evaluated on a case-by-case basis with tailored strategies for pain control. If the cause of pain is not identified after a careful review of the history, physical examination, and diagnostic evaluation, non-pharmacological methods should be considered part of the initial therapeutic plan. World Health Organization (WHO) guidelines for pain relief recommend use of pharmacologic therapies as well as non-pharmacologic measures to assist in pain control for first-line management [12]. Hypnosis (Chap. 21) and brief counseling (Chap. 18) can be very useful in the pediatric emergency department setting for alleviating pain and anxiety. Additionally, age-appropriate distraction techniques can be helpful, such as use of pacifiers for newborns, toys with lights or bubbles for toddlers, video/books for school age children, and video games for teens.

Chest Pain

Chest pain is a common and usually benign complaint in children that causes undue concern in their caretakers as they often may assume that there is a cardiac etiology (Chap. 3). Given the symptom complex, when a child presents to the emergency department complaining of chest pain, sometimes a full, thorough evaluation needs to be completed. Pediatric chest pain can be caused by many processes including respiratory, cardiovascular, gastrointestinal, musculoskeletal, neurologic, trauma, or psychogenic categories.

Psychogenic causes account for approximately 30 % of pediatric chest pain cases. Anxiety following a stressful event is the most common psychogenic trigger that induces chest pain in children [13]. Often these children have other somatic complaints in addition to their chest pain, such as headaches or abdominal pain and most have accompanied sleep disturbances. Psychogenic chest pain can mimic chest pain of more serious etiologies and can lead to extensive, unnecessary testing and potential side effects from those tests.

Pulmonary etiologies likely to cause chest pain include pneumonia, pulmonary embolism, pulmonary effusion, asthma, and acute chest syndrome (in individuals with sickle cell anemia). A chest X-ray is often performed for any child with chest pain in the ED to rule out parenchymal lung disease.

Cardiac causes of chest pain can include pericarditis, myocarditis, coronary artery pathology, arrhythmias, angina, and infrequently ischemic coronary disease. The workup to rule out cardiac causes in the ED can include an electrocardiogram, echocardiogram, and further laboratory testing. The potential for cardiac disease also induces a significant amount of emotional and psychological stress for the family. Thankfully, cardiac etiologies as the main reason for pediatric chest pain are very rare (0.3 %) [14].

Some common gastrointestinal causes of chest pain include gastroesophageal reflux disease, gastritis, esophageal spasm, dysphasia/esophagitis, gastrointestinal obstruction, or gastric ulcer. Evaluation for gastrointestinal diseases commonly includes trials of acid suppressant medications or dye imaging modalities. In the ED, a child is usually given a trial of oral medication and observed for symptom resolution.

There may be an element of excessive gastric acid secretion in a patient with a functional disorder due to persistent anxiety or stress. Stress ulcers may occur and benefit from acid blockade and mucosal coating. However, even after these measures are instituted, patients may report no significant change in their symptoms.

Management and Treatment

Management of a child with chest pain can pose a diagnostic and therapeutic challenge of critical importance. Further, functional symptoms may complicate the diagnostic evaluation. For example, when a child is stressed or anxious, he/she may hyperventilate, which can result in the development of chest pain. Hyperventilation is frequently accompanied by paresthesia and lightheadedness. Increased respirations can lead to more air swallowing and gastric distension that irritates the diaphragm leading to persistent hyperventilation. If prolonged hyperventilation leads to hypocapneic alkalosis, this can cause a vicious cycle of increasing sense of doom and anxiety that results in more rapid respiration and potential syncope. Any of the above-mentioned symptoms (chest pain, paresthesia, lightheadedness, or syncope) may trigger a large evaluation and workup in the ED. For example, chest pain that is sudden in onset may warrant an investigation for a myocardial ischemic event or a pulmonary embolism. Both of these diagnoses require immediate attention and can be serious if not recognized.

As chest pain has a vast differential diagnosis, rather than starting with an invasive workup, the medical provider may begin with alternative therapies (hypnosis, cognitive behavioral therapy, calming techniques, and distraction methods) that can redirect the attention of the patient away from the pain. In many cases, pain and anxiety relief can occur via a patient's ability to withdraw from the high intensity, often overstimulated ED setting. Redirecting a child's thoughts and calming techniques can be useful tools for children that provide relief in addition to medicine.

A study evaluating the efficacy of hypnosis for noncardiac chest pain randomized participants (adults and children) to hypnotherapy versus supportive therapy and a placebo medication. The study demonstrated greater improvement in overall well-being in addition to reduction in the use of required medication in the hypnotherapy group [15]. When pain relief is not achieved adequately with non-pharmacological treatment, analgesic or anxiolytic drugs should be considered.

If the pain, regardless of the cause—functional or otherwise—disturbs the child's activities of daily living, relieving pain with medicines is important. As the first-line therapy, acetaminophen and ibuprofen are recommended because of their safety profiles. If pain continues, adjuvant medications (e.g., antidepressants) may be considered in conjunction with analgesics. Other common therapies for noncardiac chest pain include analgesic relief and stretching exercises (such as for costochondritis), heating or cooling therapy (for muscle pathology), or allowing the child to lay in the position that is most comforting (to prevent splinting).

Syncope

Up to 15–20 % of children will experience at least one syncopal episode, and syncope accounts for 1 % of all pediatric ED visits [16, 17] (Chap. 3). The likelihood of identifying the etiology of a syncopal episode is low. Its etiology often is benign, as approximately 40 % of children may not have an organic etiology for their syncopal presentation [17, 18]. In one study, 6 % of patients with syncope presented with associated psychological issues such as depression or school phobia [19]. Nevertheless, syncope can be a manifestation of worrisome underlying pathology and warrants careful evaluation. Unlike in adults, in whom syncope often results from malignant cardiac arrhythmias, the etiology in children is more often secondary to psychosomatic causes [20]. However, given the likelihood for serious medical morbidity, the diagnostic evaluation can be extensive and lead to enormous expenditures. For example, a National Hospital Ambulatory medical care survey demonstrated the overutilization of CT/MRI imaging for diagnostic evaluation in children presenting with syncope [16]. Additionally, this can be problematic because of recent literature that suggests excessive radiation exposure in children can lead to increased risk of malignancy, given that the pediatric population has increased organ sensitivity to radiation, longer life expectancy, and increased future potential for radiation overdose with CT protocols [21].

Management and Treatment

The majority of syncopal presentations can be diagnosed by a good history and physical exam [17]. A thorough history including the regarding the presence of consciousness during the episode may help to establish whether the event was a true syncopal event (as opposed to other alterations in mental status). Additional historical details to obtain are the presence or absence of jerking/rhythmic limb movements, recovery time, presence of aura, triggers, or any other associated symptoms. A physical exam can further delineate and narrow the differential to nonproblematic etiologies. If, after a thorough history and physical, the differential diagnosis is still vast, the healthcare provider can then consider doing a workup to exclude cardiac (arrhythmia, heart block), neurogenic (seizure, stroke), pregnancy in menstruating females, anemia, electrolyte imbalances, and psychosomatic etiologies. Psychosomatic etiologies generally are diagnoses of exclusion.

Recurrent Abdominal Pain

About 10 % of children presenting to the ED have a chief complaint of abdominal pain [22] (Chap. 5). The etiology for abdominal pain can be as simple as gastroenteritis to a more pressing surgical emergency (e.g., intussusception, appendicitis, or bowel obstruction). A thorough history and physical with imaging may be necessary to exclude causes of abdominal pain that require emergency intervention. In the majority of cases, abdominal pain in pediatric patients usually is benign. In a retrospective chart review of 962 children presenting to an emergency department for acute abdominal pain, only 2 % had a surgical cause. The most common cause of the acute abdominal pain (~48 %) was constipation [23]. Functional abdominal pain (FAP) is another common cause of abdominal pain in children. According to Rome-III criteria, FAP occurs at a minimum of three times in a 3-month period without an identifiable organic etiology [24]. Its prevalence is up to 12 % of the general population.

FAP in children can interfere with their ability to perform regular daily activities and thus decreases quality of life [25, 26].

Management and Treatment

The American Academy of Pediatrics (AAP) subcommittee on chronic abdominal pain believes the diagnosis of chronic abdominal pain in children 4–18 years of age can be made by thorough history and physical examination alone [25]. Comprehensive treatment of pediatric abdominal pain requires identification and, if possible, reversal of physical and psychological stress factors that may play a role in the onset, severity, exacerbation, or maintenance of pain [27]. The AAP recommends evaluating and treating chronic abdominal pain in the context of a biopsychosocial model of care. It is imperative to address psychosocial factors in the diagnostic evaluation and management of these children [25]. A randomized controlled trial assessing medical therapies plus cognitive behavioral therapy reported significantly less abdominal pain as compared with medical therapy alone [28].

Many studies of children with FAP have demonstrated the extent that these children experience difficulties with psychological functioning. Children with FAP have elevated levels of anxiety and depression [29]. Therefore, FAP may co-occur with anxiety. This association may be due to underlying heightened physiological arousal or as a consequence of prolonged coping with pain. Screening for anxiety in the ED should be an important diagnostic consideration in the routine evaluation of a child with recurrent abdominal pain [30].

Thus, psychological treatment is recommended when addressing recurrent abdominal pain. It is imperative that medical providers explain to the patient that serious organic causes have been ruled out and that proper coping skills are necessary. Hopefully, referral for a blend of cognitive and behavioral strategies will help to improve function, reduce pain, and improve the child's ability to cope with the pain. Possible pharmacological interventions include antidepressants, antacids, antispasmotics, and prokinetics. However, the AAP and the North American Society for Pediatric Gastroenterology, Hepatology and Nutrition (NASPGHN) society both have found limited evidence to support pharmacologic therapies [31].

Fatigue

Fatigue can either be isolated problem or secondary to a previous health condition. For example, adolescents who contract infectious mononucleosis are fatigued, but once they recover from their viral illness the fatigue should improve as well. Unfortunately, sometimes even when the child/teen has recovered from infectious mononucleosis, he/she still seems to have exaggerated fatigue, which can be a result of underlying depression or anxiety.

Undiagnosed depression can cause fatigue of significant degree in adolescents. Depression also routinely causes diminished appetite, difficulty in focusing, lack of productivity at school and home, and disinterest in previous hobbies. These derangements from the adolescent's baseline can strike fear in the parents and lead to a visit in the local ED. Malignancy, chronic inflammatory disorders, irregular sleep patterns, anemia, pregnancy, and immunodeficiencies are also known to cause fatigue. These systemic processes can also cause changes appetite and weight, and decreased sense of well-being. These more chronic, serious etiologies can induce a sense of worry and disability that may exacerbate the perception of poor health.

Management and Treatment

In the initial ED workup for fatigue, lab work and perhaps imaging studies should be done to exclude any underlying pathology. Lab evaluation generally includes a complete blood count, basic metabolic panel, pregnancy test in menstruating females, thyroid studies, and possible imaging of the head. If a thorough history, physical examination, and diagnostic studies are non-revealing, the patient likely has psychogenic reasons for the

chronic fatigue. In the ED, fatigue due to psychogenic causes is managed by referral to an outpatient-counseling agency for the patient. In addition, coping mechanisms can be taught and their usage is encouraged. Close follow-up with an outpatient provider is established and then the patient is discharged.

Case Studies

Case 1

A previously healthy 16-year-old female presented to the pediatric emergency department via Emergency Medical Services because of acute onset chest pain that developed just before a 5-km run. She was the daughter of professional parents who expressed very high expectations of her and had been maintaining a near perfect grade point average at school. She had been attending a summer camp for elite teens, where she was in an environment where she was no longer the smartest child. She had struggled to excel during competitions and had expressed worry that this information would be relayed to her parents. She said she was worried her parents would think less of her because she was no longer the "smartest" child. When she developed her pain she sat down on the ground and put her hand to her left chest. She reported a sharp pain with radiation down her left arm that prompted a call to 911. In the ED, she stated that her chest pain persisted, radiated down her left arm, and was associated with finger numbness and tingling. She also complained of associated palpitations, lip numbness, finger tingling, and shortness of breath without any evidence of trauma, recent illness, intoxication, drug use, or sick contacts. Her physical examination was normal, and her pain was not reproducible by pressing on her costochondral joints during the physical examination. She underwent basic blood work, an electrocardiogram, and a chest X-ray. All results were normal. When asked how the staff could contact her parents, the teen became very diaphoretic and anxious. She begged for her parents not to be notified. She cried that if her parents knew she was in the ED, they would withdraw her from the summer camp that she had dreamed of attending for years. Suddenly, she began to hyperventilate and began complaining of worsening chest pain.

1. What is the most likely diagnosis?
   (a) Acute myocardial ischemia
   (b) Costochondritis
   (c) Anxiety
   (d) Tachyarrhythmia
   (e) Lung parenchymal disease
2. Which is the next best step?
   (a) Counseling
   (b) Troponin-I level
   (c) Aspirin and nitroglycerin
   (d) CT scan of the chest
   (e) Antibiotics
3. What should the ED physician do with regards to obtaining consent for treatment?
   (a) Call her parents, regardless of her hesitation
   (b) Further investigate the hesitation about having her parents called
   (c) Treat her as an emancipated minor without parental consent
   (d) Consult social work
   (e) Ignore the need for consent

1. (c): Given the fact that this teenager was previously healthy and developed chest pain that appeared to be related to psychological stress, the most likely diagnosis is anxiety. Her preliminary ED evaluation (labs, imaging, and EKG) was normal, so the likelihood of an acute MI, lung disease, or tachyarrhythmia is unlikely. The fact that her pain was not reproducible makes the likelihood of costochondritis less likely.
2. (a): The next step for an anxious teenager who has a normal physical exam and a negative electrocardiogram and chest X-ray would be to provide her with counseling. Ordering excessive imaging and lab work would only increase her level of anxiety.
3. (b): A teenager who presents to the ED who cries when her parents are mentioned warrants a discussion about why she is hesitant to have her parents contacted. Once this is thoroughly investigated and the concerns are addressed, then it would be imperative that the parents be notified of her ED visit.

Case 2

A previously healthy 6-year-old female presents with abdominal pain. The family denies trauma, infectious symptoms, sick contacts, recent travel, or ingestions. The pain is inconsistent, dull, and located in the periumbilical region. The mother states that the child's pain began when the father left the home a few months ago after a bitter divorce. When the patient is distracted by the television, her abdominal exam resolves. In the ED, the patient undergoes a complete blood count, comprehensive metabolic panel, amylase, lipase, urine analysis/gram stain, and abdominal X-ray. All the results are normal. The patient is given analgesics without relief. After a completely negative evaluation and normal exam, the child is discharged home with close follow-up at her regular doctor's office.

1. What is the most likely diagnosis?
   (a) Appendicitis
   (b) Constipation
   (c) Small bowel obstruction
   (d) Referred pain
   (e) Psychosomatic pain
2. Which of the following laboratory investigations probably was unnecessary in this case?
   (a) Complete blood count
   (b) Comprehensive metabolic panel and amylase
   (c) Urinalysis
   (d) Abdominal X-ray
   (e) All of the above were unnecessary
3. Which of the following is most likely contributing to her nonspecific abdominal pain?
   (a) The parents' divorce
   (b) Unknown trauma
   (c) Child abuse
   (d) Occult infection
   (e) Malignancy

1. (e): The most likely diagnosis given this child's nonspecific history with a normal exam and preliminary evaluation is psychosomatic pain. The location, quality, and characteristics of the pain make it unlikely to be an acute surgical cause (appendicitis or small bowel obstruction). Constipation is always possible in a 6-year-old female, but generally this is associated with an abnormal abdominal X-ray. It is unlikely to be referred pain because she has no other source.
2. (e): All of the above tests are unnecessary, especially in a primary care setting, according the AAP guidelines for evaluation of children with chronic abdominal pain. However, in the ED such investigations are often performed given that the patient does not have an ongoing relationship with the ED staff.
3. (a): The most likely factor to be contributing to her nonspecific abdominal pain is the parents' divorce. Given the negative workup, the likelihood of a more pathological reason is unlikely. The next best step would be to offer this family an opportunity to receive counseling. The child's presentation could be a manifestation of attention seeking behavior, conscious or subconscious, due to the recent stressor of her parental separation.

Case 3

A 17-year-old generally healthy male presents with his siblings and parents to the pediatric ED because of persistent fatigue. His parents state that for the past month he has come home from school and slept most of the afternoon. He refuses to hang out with his friends because he always complains of being tired. He has little energy and little to no appetite. He has lost a significant amount of weight. His academic performance has declined and he is now at risk of not being promoted to the next academic year. His parents deny any fevers, respiratory, cardiac, gastrointestinal, musculoskeletal, or neurologic symptoms. Outside of his family's presence, when the patient is asked about his social life, he states that his body is weak and will not allow him to do anything anymore. When asked why he thought he was in such a condition, he stated that ever since his best friend committed suicide by hanging himself, he has been unable to find motivation for living.

1. What is the most likely diagnosis?
   (a) Depressive symptoms
   (b) Malignancy

(c) Inflammatory disorder
(d) Schizoaffective disorder
(e) Suicidal ideation

2. Which is the next best step?
   (a) Counseling
   (b) Inpatient psychiatric rehabilitation
   (c) Eating disorder therapy
   (d) Fluoxetine
   (e) Encourage the family to provide fun activities that he's always enjoyed
3. What is an important issue to address prior to discharge?
   (a) The extracurricular activities in which he engages
   (b) His social network
   (c) His ability to tolerate oral intake
   (d) His religious affiliations
   (e) If there is an intention to harm himself

1. (a): The most likely diagnosis at this time is depression, which likely arose in reaction to the suicide of the patient's best friend. He manifests many of the symptoms of depression including increase in sleep, decrease in interest, decrease in appetite, decrease in ability to concentrate, unintentional weight loss, and loss of motivation for living.
2. (a): The next best step would be to provide counseling. Suicide or another cause of death of a best friend can be a very involved and complicated setting for a teenager to comprehend. Counseling could provide a necessary outlet for this young man to express his thoughts, fears, feelings, and disinterest in life.
3. (e): The most important matter to address prior to discharge from the ED would be to ensure that this individual is not a threat to himself or others. If he is in need of supervision because there is a threat of harm, it can only be done as an inpatient with a 24/7 watch.

Conclusions

Children who present to the ED may have a multitude of complaints that are attributable to organic and/or functional issues. In order to be able to exclude the most worrisome of diagnoses, a thorough history and physical is often required. When the preliminary workup is exhausted with no revealing results, the possibility of a benign etiology should be considered. Although recurrent somatic complaints are commonly encountered in children, occult organic disease is rarely present in these cases [32]. Psychosocial issues should be considered for patients with recurrent ED visits for the same presenting complaint [2]. Alternative medical therapies should be considered as part of the treatment regimen for patients with chronic pain, chest pain, syncope, abdominal pain, and fatigue that are thought to have arisen in association with functional issues.

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Functional Symptoms in Intensive Care: The Vicious Circle of Anxiety and Symptoms

16

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Abstract

This chapter reviews the functional symptoms likely to be encountered in the pediatric critical care setting. Considering symptoms to be "functional" implies that a psychological or emotional state contributes to the symptom. Thus functional symptoms overlap with related terms such as "psychogenic," "psychosomatic," and "nonorganic" symptoms. Functional symptoms occur on a daily basis in the intensive care unit (ICU) when a child's anxiety intensifies unpleasant symptoms. At the same time, unpleasant symptoms amplify anxiety. And if there is a vicious circle of anxiety and symptoms for the child, this is also exacerbated by a vicious circle of anxiety among the child, family, and staff. Symptoms during routine care, procedures, or crises in the ICU can be controlled by a combination of pharmacological and nonpharmacological strategies. Functional symptoms may mimic a life threatening condition, on rare occasions, requiring a timely diagnostic evaluation.

Keywords

Anxiety • Preparation • Distraction • Reassurance

Anxiety Worsens Symptoms in Critically Ill Children

Life-threatening disorders expose the patient to an unfamiliar ICU environment, invasive interventions, and unpleasant experiences that are likely to frighten anyone. It should be anticipated that events in the ICU intensify normal age-appropriate childhood anxieties. Children of all ages fear pain and bodily harm. Older infants and toddlers are fearful of strangers, separation from parents/caregivers, and unfamiliar situations. Children at preschool ages may be afraid of

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separation from home and family and the possibility of harm occurring to parents or caregivers. Older children and adolescents tend to feel anxiety about school performance and absence from school or athletic activities, social competence, and separation from peers, independence, privacy, and death. When stressed, children often regress to anxieties and behaviors typical for a younger developmental level [1]. If an accident involves other family members, the child must cope with this stress as well as his/her own injuries. If illness, sleep deprivation, and pharmacological factors alter judgment, even adult patients may have the delusional belief that ICU staff intends to cause them harm [2].

The experience of symptoms is subjective. Assessment of symptoms is most difficult when patients cannot verbally report their severity, whether as a result of sedation, intubation, or the communication limitations inherent to an immature developmental level [3]. Unresponsive patients are not necessarily unaware of symptoms [4]. Greater intensity of unrelieved anxiety and symptoms is likely to be associated with longer lasting unpleasant memories and the potential for disabling behavioral consequences. For children with chronic disease who will need future invasive procedures, unpleasant early experiences often result in greater subsequent anxiety, the perception that subsequent symptoms are worse, and behavioral resistance to later therapeutic procedures throughout the long course of illness [5].

Anxiety and the unpleasant experience of symptoms are interrelated as a vicious circle, each intensifying the other. Anxiety with the expectation of impending severe pain worsens the subsequent experience of pain [6–9]. In critical illness, symptoms coexist with anxiety, as pain, dyspnea, and nausea all threaten to overwhelm the patient. Inadequately controlled symptoms are likely to result in agitated behavior. Uncontrolled agitated movement risks dislodgment of, or local trauma by, life support equipment. Agitation stimulates forced breathing maneuvers that may cause dynamic worsening of airway obstruction. Forced inspiration lowers intratracheal pressure, worsening upper airway obstruction. Forced expiratory effort raises pleural pressure, worsening intrathoracic airway obstruction. In croup or asthma, agitation may result in deterioration of compensated respiratory distress, progressing to failure, requiring intubation, and assisted ventilation. Agitation is likely to exacerbate systemic, pulmonary, and intracranial hypertension and impede venous return to the heart. Agitated movement increases metabolic demands to a level that may exceed impaired cardiorespiratory and nutritional capacity. If inadequately controlled, pain also results in a shallow splinting respiratory pattern with failure to clear secretions, atelectasis, and pneumonia. Because emotional states intensify symptoms, which intensify pathophysiology, a wide spectrum of disorders in critically ill or injured patients should be regarded, in part, as functional and avoidable.

Controlling Functional Symptoms

Controlling functional symptoms requires coordinated efforts of the entire ICU team, using combinations of pharmacological and nonpharmacological treatments.

The Family

The child's family is a crucial support to children of all ages during a critical illness. In this discussion, "the family" is meant to include all those considered to be family by the child. These are the individuals who will help the child in coping with the experience of unpleasant symptoms. These may include one or both parents, members of the extended family, or a familiar and trusted caregiver. Suggestions included in this chapter focus on reducing the child's distress. Helping the family members cope with their own anxiety will promote their ability to help the child. The family's ability to help the child cope may be intuitive for some. In other cases, the ICU staff may be able to coach a family member to be more supportive of the child. Support for all family members is an important responsibility for the ICU team, detailed in standard references [10].

The Environment and Expectations

Control of functional symptoms begins immediately at the time of ICU admission as the ICU team creates a therapeutic and reassuring environment. Admission to the ICU is often precipitated by a life-threatening crisis and is a time of great anxiety for the child and the family. The child and family should quickly understand, from observing the actions of staff, that the ICU is a calm and caring environment, and that the child is under the care of an attentive, experienced, and confident team.

The child's and family's anxiety should be reduced by establishing reassuring and realistic expectations. This is communicated verbally and by example: (1) "We will be there for the child constantly," (2) "We will always act to relieve discomfort," (3) "We will always be there to adjust treatment, whenever necessary," (4) "We encourage you to stay with your child," and (5) "We will keep you informed of the child's condition and our recommendations for further evaluation and treatment." When appropriate to the situation, the family should understand that all our efforts in the ICU are intended to restore the child to a healthy condition. In some cases, the family and providers have already decided that end-of-life care or palliation is the therapeutic goal.

Unlike ambulatory care encounters where the child and family are likely to meet only one or a few providers during a brief visit, an inpatient stay involves interaction with a large interdisciplinary team. Changes of shift introduce a constant sequence of unfamiliar staff. It is inevitable that patients and family may hear subtle inconsistencies in messages from various team members. However, all staff must reinforce the basic expectation: Continuity of ICU care is in the hands of an experienced team.

The Critically Ill or Injured Child

The intubated, sedated child who may have a brain injury, and who may be treated with neuromuscular blockers, appears unresponsive. It is difficult to determine the child's level of awareness or how much the patient will remember disturbing ICU experiences. The ICU patient who appears unresponsive should be treated in an age-appropriate way as though the child can hear and understand. All patients should be spoken to in a reassuring way, again and again, providing orientation to where the child is, what time and day it is, why the patient is here, why the patient may not be able to speak with an endotracheal tube in place, or why the patient cannot move during administration of muscle relaxant. Bowel and bladder function is a concern to children at any age. The presence of a bladder catheter and the temporary use of a diaper should be explained. The patient should be warned ahead of time for uncomfortable procedures such as endotracheal tube suctioning or blood drawing and provided with a supplemental dose of analgesia when needed.

It should be anticipated that children are anxious in an unfamiliar environment like the ICU. As soon as circumstances allow, ICU staff should begin normalizing the child's experience of the environment. This includes allowing the family to stay with the child as much as possible and participate in appropriate aspects of the child's care. ICU care should be clustered together in time to allow other uninterrupted periods experienced as safe and free of frightening stimuli. When possible, it is helpful to the child to have a predictable routine of meals, play, and sleep. For older children and adolescents, privacy, independence, and social interaction become priorities for the patient, once life threatening crises have been controlled. When appropriate, allow the older child to have control of reasonable choices in daily routines. Boredom during the subacute phase of the ICU stay may seem like a relief after the terror of life-threatening crises. However, boredom and depression also interfere with recovery. Movies, games, music, stories, and visitors all may be helpful during the ICU recovery phase.

Analgesics, Sedatives, and Other Pharmacological Treatments

Since the subjective experience of unpleasant symptoms may be relieved or intensified by expectations, it is important that reassurance is quickly accompanied by pharmacological interventions that relieve pain, anxiety, dyspnea, and nausea. Effective relief of these symptoms early in the ICU

stay may improve the patient's confidence that symptoms will be relieved later and diminish subsequent need for pharmacological relief. Ineffective early relief of symptoms diminishes the credibility of reassurance and diminishes the subsequent effectiveness of nonpharmacological measures to relieve symptoms. The pharmacological relief of symptoms is partially detailed elsewhere in this book (Chap. 26) and more fully available from standard references [11].

Analgesics and sedatives may diminish respiratory drive. However, the relief of severe anxiety, pain, and dyspnea may result in more relaxed breathing, and analgesics may actually improve stability of the respiratory pattern [12]. The concern for analgesia- and sedation-induced respiratory depression in a high risk child does not warrant allowing untreated pain, but rather warrants closer monitoring during titrated incremental doses of analgesia and sedation.

Nonpharmacological Interventions

Nonpharmacological interventions are individualized to the patient, circumstances, and developmental age (Table 16.1) [13–23]. For children with developmental disabilities, strategies are chosen specific for the developmental level. When large discrepancies are found between a child's cognitive and emotional maturity, verbal communication and emotional support must each be tailored to the appropriate level. Strategies for preparation, distraction, and reassurance are applied constantly in routine care, as well as in relation to an uncomfortable procedure. Interventions may be most effective when pharmacological and nonpharmacological therapies are combined.

Table 16.1 Methods of nonpharmacological interventions appropriate to developmental age

Developmental age	Nonpharmacological interventions
Infant	• Holding/positioning for comfort • Rocking • Nursing/sucking • Relaxation singing/music • Parental presence
Toddler	• Blowing/popping bubbles • Cause and effect toys • Noisy toys • Holding/positioning for comfort • Comforting calm talk • Singing/music (Chap. 27) • Parental presence
Preschool	• Simple preparation • Electronics (i.e., Movie/TV/Tablet) • Books/Read to • Look and Find/Easy I spy Books • Blowing/popping bubbles • Holding/positioning for comfort • Play • Art therapy • Simple coaching through a procedure • Music (Chap. 27) • Parental presence
School age	• Preparation • Electronics (i.e., Movie/TV/Tablet) • I spy books or tubes • Positioning for comfort • Play • Art therapy • Intellect utilization • Music (Chap. 27) • Humor • Deep breathing • Imagery (Chap. 22) • Hypnosis (Chap. 21) • Acupressure (Chap. 23) • Coaching through a procedure • Parental presence
Adolescents	• Preparation with more detail • Electronics (i.e., Movie/TV/Tablet) • Positioning for comfort • Intellect utilization • Humor • Deep breathing • Art therapy • Journaling • Music (Chap. 27) • Imagery (Chap. 22) • Hypnosis (Chap. 21) • Acupressure (Chap. 23) • Meditation (Chap. 24) • Coaching through a procedure

Procedures and Crises

Support Person

It is useful to identify a specific individual who will serve as the support person for the child during procedures and crises. This may be a child life specialist, nurse, physician, or a family

member coached by a hospital staff person. The support person must be calm and speak with a calm voice during the procedure. Nonpharmacological measures require that the support person is simultaneously aware of the child's experience, the family's coping, the actions of physician and nursing staff, and the environment for cues to coping strategies. The support person is most effective when s/he is familiar with ICU routines and able to work closely with the ICU team. The support person must be flexible and prepared to substitute a backup approach if the initially chosen coping strategy proves inadequate.

Preparation for a Procedure

Preparation for a procedure ahead of time often reduces anxiety during the procedure for children of preschool through adolescent ages. Preparation includes explanations of the procedure, the reasons for the procedure, and how the child may feel. The child is given enough information to develop expectations about what the hospital staff will do, expectations that the support person will be there for the child during the procedure, and expectations for the child's behavior. Realistic expectations about the child's "job" in regard to the procedure helps some children structure their behavior.

The support person outlines the procedure in enough detail that there will be few surprises for the child. For example, in preparation for an intravenous catheter insertion, in an age-appropriate way, the child is told about the placement of a tourniquet, cleaning the skin, the needle puncture, and catheter placement, followed by removal of the needle only leaving a flexible plastic catheter in the vein. It may be helpful to show the child the flexible catheter that will not hurt when it is left in place.

Explanations must be appropriate for the child's developmental level. For preschool children, very simple nonthreatening language is used. The child might be told s/he will see a blue rubber band and feel a tight squeeze. Demonstrations might include enacting the procedure with pretend equipment on a doll. School age children are able to understand the logic of many medical situations if explained in simple language. Adolescents often benefit from explanations similar to those given to an adult and demonstrations of the actual equipment to be used. Older children should expect sensations such as "a pinch," "burning," and "a sting." If the older child asks if the procedure will hurt or be painful, honest answers avoid unfortunate surprises. Expectations about analgesia should be realistic. If possible, the child should be reassured that the support person will be there to talk him/her through the procedure.

Discussion in preparation for the procedure provides the support person with a chance to develop rapport and to anticipate appropriate support strategies. In preparatory discussions, a child who immediately focuses on a computer tablet movie may be favorably distracted by movies, stories, or guided imagery during the procedure. A child who has difficulty paying attention to the support person during preparations is not likely to be easily distracted during the procedure. Instead, watching the procedure and being coached in deep breathing during the procedure may be more effective for the child with a short attention span.

During the Procedure

Distraction may include a story or watching a movie on a television or computer tablet. The support person can help by positioning a book or computer tablet to block the child's view of the procedure. Guided imagery may reduce the child's anxiety about the procedure. A young child may be asked to talk about a favorite doll, Susie. "What is Susie's favorite thing to do? What toys does Susie like? What is her favorite dress?" At young ages, very specific direction helps the child to visualize distracting images. For older children, a favorite activity may be the focus during the procedure. Guided imagery/hypnotic suggestion is most effective if the support person knows something about the subject of interest and can help the child remember detailed images of a pleasant memory. "Tell me about

your favorite sport. What position do you play? Tell me about your favorite time playing that sport. What was the temperature and weather like? How did your uniform feel, soft, sweaty? How did the grass smell?…"

Other children would prefer to watch the procedure, thus gaining some control of the situation. In this case, the support person describes what is being done, consistent with expectations for which the child has already been prepared. One effective technique involves telling the child that there will be "a poke on three, one…two…three, take a deep breath, and blow away the hurt."

When There Is Inadequate Time to Prepare

The support person may be called to help the child in a crisis without any opportunity for preparation. The ICU team will determine administration of sedation and analgesics. The support person determines if a family member is present and whether that person is likely to be helpful. If so, the support person coaches the family member after a brief self-introduction. Guidance to the family may include statements like, "Now would be a good time to say…" The support person attempts to get the child's attention by calling his/her name, makes eye contact, reduces the child's view of a disturbing scene, and calmly explains the situation at an age-appropriate level. If the child is agitated and crying, quiet speech may be helpful, as the child pauses in order to hear what the support person is saying. The support person must be resourceful, noticing cues in the environment for supportive strategies such as a favorite teddy bear, a comforting blanket, or an opportunity to use a television character as distraction.

Functional Symptoms May Mimic Life-Threatening Conditions on Rare Occasions

Functional symptoms often have their basis in patients' anxieties. However, it is appropriate for critically ill or injured patients to be anxious. Thus, functional and organic symptoms may be difficult to distinguish. Functional disorders are usually recognized by primary care, subspecialty, and emergency providers because of their association with stressful precipitating events and lack of organic findings. However, an apparent crisis may lead to an intensive care unit (ICU) admission before the functional basis of the symptoms is recognized. Occasionally ICU admission follows the initiation of life support by another clinician, including endotracheal intubation, to stabilize an apparent crisis. In these cases, the life-threatening possibilities in the differential diagnosis must be promptly ruled out. The following problems are occasionally encountered in the ICU. Detailed diagnostic investigation of these disorders is discussed in other chapters of this book and in standard references.

Vocal Cord Dysfunction

Vocal cord dysfunction [24, 25] without an underlying organic cause may result in severe recurrent stridor or wheezing (Chap. 4). Inspiratory stridor or stertor suggest an upper airway obstruction. Fever and signs of a respiratory tract infection suggest the diagnoses of viral croup, bacterial tracheitis, or pharyngeal abscess. An aspirated foreign body located in a supraglottic position may either cause sudden severe stridor or subacute and progressive symptoms. In obtaining a history, inquiry may reveal the possibility of a traumatic or irritant injury to the upper airway. Occasionally central nervous system lesions result in pharyngeal or laryngeal motor dysfunction causing gradually progressive stridor or stertor [26]. Rarely hypocalcemic tetany causes laryngospasm and stridor [27]. Seizures [28] or complications of vagus nerve stimulators [29] may also be the occult causes of upper airway obstruction. Laryngomalacia and other congenital causes of obstruction, intrinsic or extrinsic to the airway, should be considered when symptoms begin in early infancy.

Acute crises due to wheezing suggest asthma, aspirated foreign body in the intrathoracic trachea or bronchus, or lower respiratory tract infection. Chronic wheezing may be due to a wide variety of airway, pulmonary, and cardiovascular

disorders. Although vocal cord dysfunction is most likely to cause noisy inspiratory sounds, and asthma often involves expiratory wheezing, in some cases it may be difficult to distinguish the two disorders [30, 31], especially as they can coexist. The diagnostic approach to stridor [32] and wheezing [33] is detailed in standard references [32, 33]. Radiographic, pulmonary function testing, and bronchoscopy may be warranted in some cases.

Chest Pain

Although most children complaining of chest pain have functional or mild disorders, potentially life-threatening causes must be considered [34] (Chap. 3). Pain results from pulmonary disorders such as a spontaneous or trauma-induced pneumothorax or the pleuritic pain of pneumonia. Vascular causes of chest pain include sickle cell chest syndrome [35] (Chap. 10), aortic aneurysm with Marfan's syndrome, and pulmonary embolus. The diagnosis of pulmonary embolus is being recognized more often in the pediatric age group, especially adolescents. Risk factors for pulmonary embolus include immobilization, a known hypercoagulable disorder (including a known deep venous thrombosis), oral contraceptives, and an indwelling central venous catheter [36]. In adults D-dimer assay is increasingly relied upon as a screening test to guide more definitive contrast CT scanning for pulmonary embolus. However, this screening approach in children has not yet been validated. Cardiac ischemia will seldom be found as the cause of chest pain in a child, unless an underlying risk factor is present such as prior Kawasaki Syndrome, anomalous coronary artery, aortic stenosis, hypertrophic cardiomyopathy, long-standing diabetes mellitus, or cocaine use [37]. Finally, an esophageal foreign body may be a dangerous occult cause of esophagitis, esophageal perforation, and chest pain. Radiographic assessment, electrocardiogram, and evaluation of cardiac enzymes may be warranted in selected cases.

Seizures, Focal Weakness, and Coma

Neurological symptoms may have a functional basis that must be distinguished from life threatening neurological disorders (Chap. 2). Pseudoseizures are typically chronic and recurrent, without life-threatening manifestations, unless anticonvulsant therapy causes iatrogenic respiratory depression. Pseudoseizures in children may resemble generalized tonic–clonic (60 %), focal clonic (10 %), or akinetic seizures (10 %) [38]. Pseudoseizures seldom result in incontinence, injury, tongue biting, stertor, and seldom occur while asleep [28, 38].

Acute focal weakness suggests the possibility that the patient has had a stroke. In one pediatric study, among 14 children with acute focal weakness who did not have a stroke, six were found to have a benign diagnosis, including a functional basis in two [39], while others had migraines or musculoskeletal disorders. An inconsistent pattern of weakness on repeated exams, normal movement of the involved extremity during sleep, and patterns of sensory and motor deficits that do not correspond to a neuroanatomic basis may be clues to a functional disorder.

When organic disorders result in coma, patients will typically allow an arm dropped over the face to strike the face. In functional disorders mimicking coma, the patient may pull the dropped arm away to avoid striking the face. Because so many disorders with severe consequences may cause neurological symptoms in an apparently critically ill patient, an EEG, neuroimaging, and CSF examination will often be warranted to rule out a life-threatening organic disorder before considering the diagnosis of a functional cause [40].

Professional Mental Health Evaluation and Therapy

Consider consulting a mental health provider when functional symptoms are severely disabling, and particularly when functional symptoms were the primary event precipitating a hospitalization (Chap. 18).

Brief Cases

1. A 3-year-old presents with croup, severe respiratory distress, and agitation. His $SpO_2 = 100\ \%$ in blow-by oxygen. Appropriate action would be:
   (a) Allow the child to remain in a position of comfort in parent's lap
   (b) Administer racemic epinephrine aerosol, IV dexamethasone, and inhaled helium oxygen mixture
   (c) Consider titrated incremental doses of midazolam while continuously monitoring the patient
   (d) Prepare to intubate the patient
   (e) All the above
2. A semi-alert 5-year-old in septic shock needs to have a central venous catheter inserted. Appropriate action would be:
   (a) Prepare the child ahead of time by explaining what will be done, including measures to keep him comfortable
   (b) Administer analgesic and sedative
   (c) Have the child life specialist or other staff member distract the child by guided imagery
   (d) Implement standard infection control practices
   (e) All the above.
3. A 10-year-old complains of the acute onset of unilateral weakness and loss of sensation. The weakness appears to be inconsistent on repeated exams. Appropriate actions would include:
   (a) Request a neurology consultation.
   (b) Consider an MRI scan.
   (c) Document progress on sequential exams.
   (d) When the MRI scan is normal request a psychiatry consultation.
   (e) All the above.

Answers

1. (e): In a position of comfort the child is less likely to become anxious and thus exacerbate the respiratory distress. A benzodiazepine may reduce anxiety, thus reducing the dynamic exacerbation of airway obstruction associated with forced respiratory effort. Epinephrine aerosol and dexamethasone may reduce mucosal edema, while inhaled helium may improve laminar flow in the obstructed airway thus reducing work to breath. Intubation may be necessary in progressive respiratory failure.
2. (e): Combinations of pharmacological and behavioral support are likely to be most effective.
3. (e): All of these actions are appropriate.

Video Case

The patient in Video 16.1 is a 13-year-old with velo-cardio-facial syndrome (Chap. 13) who has been hospitalized for a week as a result of an aspiration pneumonia. He has chronic lung disease related to recurrent aspirations and other medical complications, which have necessitated that he have a tracheostomy placement. However, he did not require mechanical ventilation at home before his last aspiration episode and was started on ventilation during his current hospitalization. This video demonstrates part of a session of chest physiotherapy.

1. Should the respiratory therapist have told the patient lying in bed that he was sliding away?
   (a) Yes, as this is a metaphor the patient can understand.
   (b) Yes, as she needed to give a reason for having to readjust his position.
   (c) No. The patient might have misinterpreted the comment, and felt uncomfortable, especially if he has had a bad experience involving a slide in the past.
   (d) A & B
   (e) None of the above.
2. Should the respiratory therapist give the patient a choice regarding which side should first be administered chest physiotherapy?
   (a) Yes, as this allowed the patient to maintain a sense of control in an ICU setting.
   (b) Yes, because giving the patient a choice demonstrated respect for his feelings.
   (c) No, because he really did not have a choice, and it is unwise to give a false choice.

(d) A & B
(e) None of the above.

3. Was it important for the respiratory therapist to have told the patient he was doing "awesome?"
   (a) Yes, as children like to be told they are doing well, even if it is not true.
   (b) Yes. It helped him feel as he was helping with the procedure.
   (c) No, because it was just a throw away word that was uttered to fill the time.
   (d) A & B
   (e) None of the above.
4. When the respiratory therapist wanted the patient to help turn himself, which of the following suggestions would have been most helpful to prompt the patient?
   (a) Do you feel tired today?
   (b) Are you being lazy today?
   (c) I've seen you turn yourself. Can you show me how you do it?
   (d) A & B
   (e) None of the above.
5. How is the patient likely to react when the respiratory therapist asks the patient to try to keep his arm down?
   (a) Keep his arm down.
   (b) Make a concerted effort to cooperate.
   (c) He is more likely to flail his arm.
   (d) A & B
   (e) None of the above.
6. Was it helpful for the patient to be asked whether it was "OK" for the chest physiotherapy to be administered at the "last spot?"
   (a) Yes, because this question showed respect for the patient's autonomy.
   (b) Yes, because it showed the therapist cared about the patient's feelings.
   (c) No, because when the patient refused, the chest physiotherapy nonetheless was administered.
   (d) A & B
   (e) None of the above

Answers

1. C: While the patient should have been told what is going to happen, using a metaphor that could be misinterpreted by a young child can lead to an uncomfortable reaction. Instead, the respiratory therapist might have told him that she is going to help him feel more comfortable by repositioning him.
2. D: The patient was not asked if he desired chest physiotherapy, to which he might have replied negatively, and then his wishes and autonomy would have been overridden. Instead, the respiratory therapist gave him a choice of which side to first use, which maintained his autonomy while providing information that he will be receiving chest physiotherapy.
3. B: Children know when people are nonauthentic. Therefore, healthcare providers should identify truthful observations that help their patients feel better. Words can have a big impact on patients and should not be used without thought as to their impact.
4. C: A positive suggestion is more likely to yield a cooperative patient. When it is suggested that the patient might be tired or lazy this serves to reinforce the patient's lack of cooperation rather than as a motivator for a change in his behavior.
5. C: When the therapist suggested "trying" to keep the arm down, the patient could move his arm, while thinking to himself that he was cooperative because he "tried." A better suggestion would have been, "Keep your arm down as best as you can." Notice how the patient responded shortly thereafter when it was suggested, "Do you want to put your arm down?"
6. C: When the patient's negative response was disregarded, this represented a disruption of the relationship with the therapist and was harmful to the patient's feelings of autonomy. A better question might have been, "Do you want me to start chest PT now so we'll be done quickly, or do you want to take a break for a couple of moments?"

Conclusions

Considering symptoms to be "functional" implies that a psychological or emotional state contributes to the symptom. Functional symptoms occur on a daily basis in the ICU. Symptoms during routine care, procedures, or crises can be controlled by a

combination of pharmacological and nonpharmacological strategies. Functional symptoms may mimic a life-threatening condition, on rare occasions, requiring a timely diagnostic evaluation.

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Development of Functional Symptoms in Children Exposed to Traumatic Events

17

Paula A. Madrid and Robert K. Kanter

Abstract

This chapter will review the typical symptoms occurring in children after stressful traumatic exposures. Unlike other chapters in this book, no specific organ system is the most likely focus of functional symptoms in this setting. Psychological distress may exacerbate symptoms of physical illness and injury associated with the traumatic events, may be expressed as almost any seemingly unrelated symptom, may intensify the age appropriate fears typical of any child, or may predominantly be exhibited behaviorally. In most nonsevere cases, the impact is self-limited and the individual's functioning will be back to normal within days or weeks. We will suggest simple behavioral and environmental interventions intended to help relieve children's distress. However, when large populations are affected and individuals suffer severe loss such as in a mass casualty disaster, the scale of events requires community-wide efforts to meet the needs of children and their families. The fact that some children are more psychosocially vulnerable than others will be discussed. The chapter will conclude by highlighting warning signs warranting professional mental health care.

Keywords

Children • Trauma • Behavior • Mental health • Disaster • Mass casualty

Introduction

When physical or emotional harm occurs to oneself, a loved one, or even when witnessing harm to a stranger, most people experience the event as emotionally upsetting. Likewise, the loss of important possessions and major disruptions of routines can be distressing. Stressful exposure to traumatic events is an inevitable consequence of human existence. Such stress may occur as a result

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of accidental or intentional violence involving individuals or affecting a large population as in mass casualty disasters.

Child Anxiety

All children exhibit some form of anxiety according to their developmental stage. Children fear pain and bodily harm at all ages. Older infants and toddlers are fearful of strangers, absence of parents/caregivers, and unfamiliar situations. Children at preschool ages may be afraid of the dark, animals, separation from home and family, and the possibility of harm occurring to parents or caregivers. Older children and adolescents often feel anxiety about school performance, social competence, independence, privacy, and death. Traumatic events are experienced as threats that intensify usual age appropriate childhood anxiety. When stressed, children often regress to anxieties and behaviors typical for a younger developmental level [1]. The anxiety manifested as a result of exposure to trauma intensifies age-appropriate childhood anxieties.

Epidemiology of Stress Responses: Everyday Traumatic Events

Most children experience a traumatic event at some time, and these events are often brought to the attention of a primary care physician. Thus primary care providers are in an ideal position to screen and provide preliminary support for children who might otherwise remain without help. Likewise, emergency physicians often have the opportunity and responsibility to identify and help children with mental health emergencies [2].

Children's stress reactions tend to be proportional to the suddenness of onset, the severity of life threat, injury, exposure to the grotesque, separation from caregivers, physical losses, and the severity of environmental disruption [3]. For instance, after being directly involved in a motor vehicle accident, 11 % of adolescents reported subsequent stress-related symptoms (Table 17.1), fear, and depression. These symptoms exceeded stress symptoms in a control population. Adolescents who witnessed a motor vehicle accident also had an increased risk of such symptoms even when they were not directly involved [4, 5]. Fourth-grade children who witnessed the accidental deaths of two adults at a school were so upset that many could not attend school for several days. Then symptoms of stress rapidly declined in subsequent days [6]. Traumatic events occurring within a family, whether physical or sexual abuse, emotional maltreatment, neglect, sibling victimization, or witnessing family violence, are especially likely to cause stress symptoms [7].

Table 17.1 Common early symptoms

Emotional reactions
Fear
Shock
Disbelief
Feeling overwhelmed
Feeling hopeless
Feeling numb
Panic attacks
Behavioral reactions
Sleep disturbance
Hypervigilance
Irritability
Exaggerated startle
Aggressive outbursts
Apathy
Withdrawal from interactions
Separation anxiety
Regression to less mature behavior
Cognitive symptoms
Difficulty concentrating
Difficulty making decisions
Poor attention span
Loss of memory
Preoccupation with unpleasant memories
Physical symptoms
Gastrointestinal distress
Headache
Nausea
Dizziness
Tachycardia
Tremor
Generalized pain
Fatigue

Epidemiology of Stress Responses: Disasters

Disasters involving large numbers of illnesses, injuries, deaths, and community disruption trigger similar stress responses as everyday traumatic events. However, because of the widespread exposure to distressing events and disruption of therapeutic services, the impact may be exaggerated and last longer. Even when a public health emergency caused few life-threatening implications for the population, community disruption and environmental hazards resulted in one-third of parents reporting physical symptoms or mental health issues that affected their children after the 2010 Gulf Coast oil spill [8]. Likewise, as measured by the proportion of children receiving counseling, 22 % of children in Manhattan experienced significant stress-related symptoms in the aftermath of the September 11, 2001 attack [9]. Media coverage of emergencies is often intense, so that exposure to the disturbing events is repetitive. It has been found that media coverage may provoke stress responses in individuals remote from the events who suffered no direct involvement to themselves or loved ones [10]. Thus, immediately after exposure to a traumatic event, virtually all children and adults may exhibit stress-related symptoms and behaviors, and this is well within the normal range of responses.

Common Early Symptoms After Traumatic Events

The early symptoms of traumatic stress include emotional reactions [11–13]. It is common to feel fear, shock, disbelief, and seemingly being overwhelmed, helpless, or numb. More serious symptoms may include dissociation and panic attacks. Common cognitive deficits include difficulty concentrating or making decisions, shortened attention span, loss of memory, or preoccupation with unpleasant memories. Physical symptoms may include gastrointestinal distress, headaches, nausea, dizziness, tachycardia, tremor, generalized pain, and fatigue. The clinician may find increased arousal manifested by increased blood pressure and heart rate. Hypervigilance, sleep disturbance, irritability, exaggerated startle response, and aggressive outbursts are common behaviors [14]. Immediately after a traumatic event, such symptoms and signs may confuse the diagnostic evaluation of physical injuries (see Table 17.1).

After traumatic events, individuals may be irritable and behave aggressively or may become sad, apathetic, and withdraw from interaction. For example, the mutism of many unaccompanied injured children immediately after a tornado interfered with medical history taking, and delayed family reunification [15]. Age-specific trends may be noted. Toddlers and young children are likely to demonstrate increased crying, separation anxiety, clingy behavior, sleep terrors, nightmares, and tantrums. School age children display aggressive or thematic play (reenactments, ritualistic interactions and a fascination with a particular person, toy, or character), specific phobias, sleep disturbance, physical complaints, withdrawal, regressed behavior including enuresis, thumb-sucking, clinginess, baby talk, asking for a bottle, and separation anxiety. Adolescents may be depressed, anxious, withdrawn, do poorly in school, be irritable, deny that a permanent change has taken place, or engage in risk-taking behavior. Adolescents may attempt to mask symptoms, fearing that their reactions might seem abnormal. For the majority of children, as long as they do not experience further stressors, such symptoms usually diminish over days or a few weeks.

Simple Early Interventions for Children Exposed to Traumatic Events

Any emergency care, primary care, school health, or subspecialist provider may be able to help children and their families by giving simple advice in the immediate aftermath of a traumatic event. Family collaboration is a must whenever possible.

– Reunify the child with family members at the earliest possible time, when relative is not the perpetrator of trauma.

- Provide transitional toys, safety blankets, or other coping toys to younger children.
- Reassure the child or family member honestly that "keeping you safe is the most important thing to me," and "we will do everything we can to find your parents."
- Give the child realistic, age-appropriate hope.
- Maintain familiar routines whenever possible (family, school, church).
- Identify concrete needs and try to help meet them.
- Listen to the child. The amount of information given to the child depends on what s/he wants, for which s/he is ready.
- Do give children information about the event. Be aware of information the child is receiving on television, the Internet, and what s/he has actually seen on the street.
- Encourage children to express feelings in words, play, behavior, and drawings or other art forms.
- Keep in mind that children's responses may be confusing and confused. Denial does not mean the child is not distressed or unaware.
- Children are resilient if supported by adults. Adults who model effective coping behavior promote healthy coping skills by children [16].

Organizational Approach to Mental Health Services After Disasters

A surge in population needs for mental health services may be anticipated when large numbers have experienced risk and traumatic events, especially if accompanied by restriction in their movement, limitation in resources, and inadequate information [17]. Restoring communications systems will reduce the sense of isolation, provide the public with information to avoid hazards, and promote population safety, as well as relieve anxiety associated with separation from loved ones. Child safety, identification, and reunification procedures improve efficiency and effectiveness of all other services [18].

School resources have been vital in supporting community-wide early efforts to help children and families cope with the traumatic experiences of a disaster [16, 19–21]. Important opportunities include early availability of the school as a safe environment that restores familiar routines. School administrative resources may be useful in contacting all families and staff to determine needs and to coordinate ongoing assistance with housing, childcare, and health care. School-based group counseling may be an efficient way of providing simple interventions for large numbers of children. School-based screening also identifies those whose greater needs warrant referral for professional mental health care (see below). "Just-in-time training," brief educational sessions focused on a specific set of problems and solutions, optimizes the role of schoolteachers in helping children, and recognizing severe mental health problems.

Vulnerability for Severe Mental Health Disorders After Traumatic Events

Children with the most severe traumatic experiences are at greatest risk for severe and prolonged mental health disorders, whatever the prior individual, family, or neighborhood factors predisposing to vulnerability or resilience [3, 22]. Greater threat to life or actual loss of life of a loved one, injury, exposure to the grotesque, victimization due to lawlessness, separation from caregivers, and physical losses all raise the risk of prolonged and severe mental health issues.

Some children's personal situations place them at greater risk of severe mental health consequences after a traumatic event. These risk factors include characteristics of the child (prior mental health problems, medically vulnerable), parents (poor mental health, low socioeconomic status), household (unstable housing, lost income, economically disadvantaged), and neighborhood (unsafe schools and community, social disorder) [23]. Children with any disabilities and special health care needs are likely to have greater risk of mental health consequences after traumatic events [24].

Also at risk may be children whose parents serve in the military. Children in military families experience family separations with the real threat

of a parent's death or disabling injury, as well as the frequent need to move to a new community. Also families must cope with the stress responses of veterans returning from combat. In the opinion of one American military leader in 2012, adolescents in military families may feel that "my whole conscious life has been at war. The U.S. has never, never experienced that before" [25].

Warning Signs of Severe Mental Health Disorders After Traumatic Events

In addition to persistence of the common stress response symptoms described above, more severe manifestations must be recognized.

Self-Injurious Behaviors

Self-injury takes many common forms such as burning, cutting, interfering with wound healing, and scratching. It can also take the form of banging body parts, needle sticking and carving words or symbols into the skin [26], gouging, hair-pulling or plucking, inserting harmful objects into the vagina or rectum, picking at cuticles and biting nails until bleeding occurs, biting oneself, digging into the gums, choking, hitting oneself with objects, ingesting sharp objects, and using an eraser to tear or burn the skin [27].

Suicide Risk

Childhood sexual abuse and neglect is a factor in increased vulnerability to suicidal ideation [28, 29] as are the presence of psychiatric disorders such as major depressive disorder, generalized anxiety disorder, panic disorder, attention-deficit hyperactivity disorder, conduct disorder, and substance use disorder [30]. Studies have also found that current alcohol consumption, use of illicit drugs, symptoms indicating conduct disorders are risk factors for suicidality for children 11–15 years of age [31]. Similarly, there are findings indicating a strong consensus that adolescent and adult men who report same-sex sexual orientations, identities, and behaviors are prone to experience trauma as a result of interactions with others that increases their suicide risk [32]. A study used regression analyses to determine whether children who experience childhood bullying behaviors are at risk for later suicide attempts and completed suicides after controlling for baseline conduct and depression symptoms. Its findings were that among boys, frequent bullying and victimization were associated with later suicide attempts and completed suicides. However, after controlling for conduct and depression symptoms, there was no identified association between bullying and suicidality. Frequent victimization among girls is associated with later suicide attempts and completed suicides, even after controlling for conduct and depression symptoms [33].

Substance Abuse

Adolescents with traumatic stress experiences are more likely to have issues with substance abuse and other risk-taking behavior [34].

Sleep Disturbance

Sleep disturbances are hallmark signs of trauma. These include nightmares, difficulty falling and remaining asleep, sleepwalking, night terrors, and bedwetting at night. Daytime functioning is also affected as a result of resultant fatigue and excessive daytime sleepiness [35].

Regression

Regression refers to a reversion to an earlier or less mature pattern of feeling or behavior. Regression is exhibited by children as a behavior that would be normally observed in an earlier stage of development. Bedwetting, clingy, whiney behavior, and engaging in baby talk are common [36]. Long lasting regression may indicate a persistent disorder warranting professional mental health evaluation.

Selective Mutism

Selective mutism is a "childhood disorder that is characterized by the persistent failure to speak in at least one specific social situation, despite the ability to speak in other situations" [37]. Children who become mute in social settings tend to be talkative around family with whom they are familiar, but at school they might respond to queries with nonverbal signals such as gestures and nods. These children are sometimes either rigid or fidgety and may blush when communication is attempted. Efforts to force them to speak will result in an irritable disposition, an outright temper tantrum, or severe withdrawal.

Cruelty to Animals

It is important to distinguish between explorative play in which children "handle" animals such as insects causing unintentional harm versus torturing pets or other small creatures. Animal cruelty, like any other violence, should never be attributed to a stage of development and if allowed to harm animals, children are more likely to be violent later in life. Children who witnessed family violence were three times more likely to abuse animals than their peers [38].

Inappropriate or Disturbing Play Behaviors

Play is a major feature of childhood activity. Effects of traumatic experiences may be expressed in play. Play behaviors predictive of Posttraumatic Stress Disorder (PTSD) include frequent acting-out morbid themes and play at an immature developmental level [39].

Poor Peer Relationships

Children who have been maltreated are usually less socially competent and may be socially rejected by their peers. Traumatized children may have difficulty establishing or maintaining relationships or reading social cues [40]. Their interactions are often aggressive, belligerent, or rebellious.

Poor School Performance

Changes in school performance should be noted, and the causes explored. Elementary school performance is impaired by stressors found in a child's school, family, and/or neighborhood. Math and reading grades decline while depression and aggression rates increase [41]. Similarly, reading performance diminished for urban 2nd through 5th graders who had experienced violence or the loss of a significant other [42]. Exposure to traumatic experiences is also a risk factor for school dropout [43].

Inappropriate Sexual Behavior

Children's sexual development begins at birth and progresses through peaks and troughs depending on their developmental stage and socialization. Some forms of sexual play are normal and expected at various ages of development. However, some sexual behaviors raise concern:

- Sexual experimentation with older children or those who are not their usual playmates.
- Sexual experimentation that results in other children making complaints.
- Undue shame, guilt, fear, or anxiety about sexual behavior.
- Child using warped reasoning to justify sexual behavior, for example, forcing another child to engage in sexual behavior and saying that the child really wanted to participate.
- Sexual interest that appears out of balance with other aspects of the child's life.
- Child engaging in persistent and extensive sexual behavior with another consenting child [44].

Sexually transmitted diseases may result from such behavior. Depending on the age of the child, pregnancy is a risk.

Separation Anxiety Disorder

Separation Anxiety Disorder (SAD) refers to an abnormal reactivity to real or imagined separation from attachment figures. It interferes with daily activities and developmental tasks [45]. This occurs at a rate of 4–5 % among children, with an average onset age of 7.5 years old. Clinical help is usually sought after the child expresses somatic complaints and/or refuses to attend school. After the 2001 World Trade Center attack [46], the prevalence was 12.3 % in school-aged children; 4th and 5th graders were three times more likely to have probable SAD than 9th through 12th graders. After the September 11 attacks, SAD was more likely for girls, for those directly exposed to the attacks, with relatives involved, and after extensive media exposure.

Posttraumatic Stress Disorder

PTSD involves a pattern of stress related symptoms including re-experiencing the stressful event repeatedly, avoidance of reminders of the experience, and hyperarousal, all lasting more than a month [12]. Repetitive experience of the event may include a vivid sense of reliving the event, nightmares, and recurrent physiological or emotional reactions to these stressful memories. Avoidance includes efforts to distance oneself from memories as well as from locations or images associated with the traumatic event. Impaired memory, feelings of detachment and estrangement, and feelings of hopeless inability to plan for the future may follow. Hyperarousal may include difficulty sleeping, angry outbursts, hypervigilance, and inappropriate startle responses. PTSD can impair home, school, and social functioning and must be distinguished from appropriate bereavement and sadness that may include recurrent thoughts, but does not unduly interfere with functioning.

Referral and Treatment

Severe symptoms interfering with function or suggesting the likelihood of self-destructive behavior are red flags. Simultaneously impaired functioning at home, school, and socially (interpersonally), as well as ongoing acute stress symptoms (longer than 1 month) call for (1) immediate psychiatric evaluation, and possibly hospitalization and (2) extended care, monitoring, or some form of ambulatory intervention [47].

Mental health professionals have a wide range of tools at their disposal for treating traumatized children.

There is support for the use of directive, trauma-focused therapy over nondirective, support-oriented techniques to reduce most child trauma symptoms [48]. Treatment approaches such as Trauma-focused Cognitive Behavioral Therapy [49] (Chap. 19), Prolonged Exposure [50] Cognitive Behavior Therapy of Childhood Sexual Abuse [49] as well as Semi-structured Interview Intervention [51], and Structured Play Therapy [52] (Chap. 28) have been widely used among many others.

It is essential to stress the importance of parents and caregivers to provide stressed and or traumatized children with a safe, consistent, and predictable environment. Immediate goals include stabilizing the home environment, making sure the threat has been removed, i.e., abusive member of the household is no longer a threat to the child and ensuring that caregivers are able to keep the child safe. Healthcare providers across disciplines must collaborate in these efforts. It is also very important for families to understand trauma and PTSD and its impact on the child's functioning. This may in turn provide the parent with the ability to normalize symptoms and reframe symptoms as signs of coping and protective healing mechanisms. It also encourages parents and caregivers to discuss and understand their own feelings about the event that caused the trauma in the child. Medical providers may also educate parents about anticipating a child's reactions during times of transition such as changes of school, holidays, and the anniversary of the traumatic event.

The psychological treatment of a child should always take place with parent/caregiver cooperation. This involves providing the caregiver with education about the child's condition and the best ways to help the child outside the therapy setting.

Sometimes, it is necessary for providers to think of themselves as "detectives" in that children of all ages can mask the source of anxiety and depression from themselves and others. In situations where children and teens refuse to participate in specific activities, perseverate in certain behavioral activities, and deny worries even when nonverbal cues indicate stress, providers are encouraged to gently pursue the "child's world." Perhaps the physical or social environment is subtly reminiscent of the trauma, and the fear has generalized to unexpected people, places, and objects.

In caring for children after exposure to traumatic events, practitioners must take into account cultural considerations such as the way in which the child and his/her family think about and cope with stress, grief and loss, the forms of acceptable assistance, and religious beliefs.

Case Studies

1. A 6-year-old boy is involved in a car accident in which he is unharmed but his parents suffer cuts and bruises. Soon after the accident, he has a difficult time falling asleep unless he is with his parents, appears to have lost his appetite, is irritable, clingy, and becomes tearful more often than before. After 2 weeks and with continuous reassurance from his parents, the boy's behavior improves and gradually returns to his prior level of functioning.
   What should the primary care provider do?
   (a) Monitor the child for additional symptomatology.
   (b) Talk to the parents about appropriate stress responses.
   (c) Properly document incident for future reference.
   (d) Inquire how the parents are doing.
   (e) All of the above.
2. A 10-year-old girl witnesses her neighbor get shot in the street one afternoon. She becomes severely shocked, refuses to sleep on her on from that point on, and her parents agree to let her sleep with them for months. She also becomes severely withdrawn, appears to be "out of it" as she is often daydreaming and loses interest in her friends, all play activities, and in school. The patient's parents are mostly concerned about her academic performance but have not yet discussed her feelings regarding the event she witnessed. The patient's teachers believe she has become lazy and disinterested in school. She finally receives counseling 9 months after the incident because her parents become concerned that she may not be promoted from 5th to 6th grade.
   What should the primary care provider do?
   (a) Encourage parents to actively participate in the girl's treatment.
   (b) Coordinate treatment with her mental health provider in case she becomes a candidate for psychiatric medication and to monitor psychosomatic complaints.
   (c) Normalize the girl's reactions by discussing common trauma response with her parents and ideally with teachers as well.
   (d) All of the above.
   (e) None of the above.
3. An 8-year-old boy's home is burned down in a fire that also destroyed four neighbors' homes. The boy was at home sleeping when firefighters save his life and that of his family. The boy and his family lose all of their possessions and are forced to move in permanently with their relatives in a nearby town. The boy is unable to return to his school and has heard that that his friend and neighbor died in the fire. His parents are so devastated by their material loss that they fail to provide him with the support, reassurance and validation he needs. Within a month, the boy has become aggressive, refuses to attend school, is throwing lit matches at his cousin's cat, has nightmares, is constantly irritable, and does not appear to care about his or others' welfare.
   What should the primary care provider do?
   (a) Immediately refer the boy to a mental health professional.
   (b) Monitor for physical symptoms and suicidal ideation.

(c) Provide referrals to parents who are also likely to have developed significant traumatic responses.
(d) Make him/herself available to discuss child with teachers and other professionals in need of insight.
(e) All of the above.

Answers

1. (e): Since the 6-year-old did not demonstrate ongoing symptoms, it is reasonable to assume that the child is doing well and will not present long-term sequelae. Nonetheless, it is important to monitor the child on the next several visits to make sure he has in fact continued to function as expected. Providers may use this opportunity to discuss normal stress responses with parents as a means to normalize and validate their concerns as well as to inquire about their own functioning. The latter is particularly important as parents' mental health status will greatly influence their children's welfare. It is also expected that providers properly document patients' traumatic exposures for future reference.
2. (d): The patient described above is in need of treatment, support, and active participation and coordination between her providers and parents. It is also essential that her parents and teachers be aware of common trauma responses and the need for long-term support and understanding of symptoms in order to prevent misjudging their child and her symptoms.
3. (e): This child is demonstrating symptoms of a severe stress reaction, possibly posttraumatic stress disorder. For this reason, it is important to immediately refer to a qualified mental health professional in order to prevent further exacerbation and to begin treatment. Traumatized children often exhibit somatic complaints and as such, the child must be monitored medically. The child's parents are also likely to be experiencing a severe stress response and as such, would benefit from an evaluation by a mental health professional who may help the family as a whole to manage their respective symptoms and needs.

Conclusions

Exposure to traumatic events results in psychological distress, whether in relation to events experienced individually or in a major public health emergency. Psychological distress may exacerbate symptoms of physical illness and injury associated with the traumatic events, may be expressed as almost any seemingly unrelated symptom, may intensify the age appropriate fears typical of any child, or may predominantly be exhibited behaviorally. Simple behavioral and environmental interventions are often sufficient to relieve children's distress. Warning signs will identify those children needing professional mental health care.

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Part II

Treatment of Functional Symptoms in Children

When and How to Refer a Patient to a Mental Healthcare Provider

18

Julie H. Linden

Abstract

Functional symptoms can at times be difficult to manage in the primary care setting. Sometimes, the concomitant psychological issues that contribute to and maintain functional symptoms require the intervention of a mental healthcare provider. In this chapter, steps are presented that can help to determine when and how to successfully refer a child or adolescent patient for this expertise. Key factors in a successful referral such as patient perspective, the constructive language of referral, and responsibility for patient care are also discussed. Effective management of functional symptoms requires consideration of a range of items. These include availability of mental health care, the cost effectiveness of treatment, the education and coaching of the physician to adequately assess the psychological issues, and to develop pertinent mental health treatment skills. Each of these items is addressed in the process of managing functional symptom presentations and finding the best treatment options for patients.

Keywords

Referral • Children • Functional symptoms • Mental health

Introduction

Functional symptoms in pediatric disease come in many forms. They are represented by the rubric of each chapter heading in this book. This chapter will focus on guidance that the primary care provider may utilize to complete the "Rubik's cube" puzzle that can accompany the management of patients with functional symptoms. (PCP will be used to refer to all pediatric physicians, family practice physicians, nurse practitioners, physician assistants, and primary care providers in any health setting for children and adolescents.)

Background

The introduction of the term functional symptoms into the lexicon of medicine has a long and interesting history [1–4] that is relevant to the practice

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of medicine [2, 5–7] and the question of if, when and to whom to refer patients for mental health care. Most agree that "functional" terminology defines a symptom as a perception of change in the body or its functions in a way that indicates disease [1]. When there are no clear medical diagnoses or treatments and when the mechanism of a symptom cannot be directly observed the symptoms often are classified as functional [7]. PCPs sometimes refer patients with functional symptoms to psychological practitioners. This contributes to the parent or patient's assumption of "something psychological" that is "causing" symptoms. This is the juncture at which referrals to mental healthcare providers typically have been considered and is the focus of this chapter. Of importance in the referral process is the complex mind/body interplay that affects the practice of medicine and mental health as well as the relational issues between patient and provider that integrative medicine strives to address.

Integrative medicine is defined as a combination of conventional mainstream medical therapies and complementary and alternative therapies, for which there is scientifically acceptable evidence of both effectiveness and safety. One of the main principals of integrative practice is the partnership between practitioner and patient in the healing process. Another is the consideration of all factors that influence health, including mind, body, spirit, and society.

Since the incorporation of the biopsychosocial [8] emphasis into health care, there has been an undoing of the dualistic notion of the mental and the physical. For example, the dismissive "It's all in his head" statement is no longer an acceptable assessment for physician or psychiatrist to utter. The old divisions between psychological and physiological processes hindered development of a holistic model [9]. From a neurobiological perspective this division is understandable. While the emphasis in medicine has been a "bottom-up" focus on the human body, its systems, diseases, and anatomy, the emphasis in psychology has been a "top-down" focus on emotions, behavior, and personality. Through employment of an integrative perspective, functional symptoms are best understood as multifaceted and multifactorial in etiology, with predisposing, precipitating, and perpetuating factors all at play [4]. Neurobiologically, functional symptoms can be viewed as occurring at the "crossroads" of bottom-up and top-down processes, below conscious awareness and fueled by physiology, and will require knowledge of both contributing processes. Integrated care and person-centered (those that value the quality of the doctor and patient relationship) models are the new emphases that challenge all healthcare providers to change old patterns of care and work collaboratively across disciplines.

What does the question that serves as the title of this chapter tell us about current medical and psychological practice? "**When** to refer a patient to a mental healthcare provider?" This statement assumes that referral will be necessary and that the PCP does not have the requisite skills or time to manage some patients or symptoms and may reinforce a belief that care will transfer in a one-way direction from medical to psychological. Each of these assumptions can interfere with the process of diagnosis and of treatment. There is quite a bit of data to support these assumptions [5, 10–14] and it is intrinsic to the structure of the medical and psychological training systems. A search of the literature in medicine or psychology produces separate "silos" of data, with few bridges between the disciplines. As both PCPs and mental healthcare providers move to an integrated model of care the questions each group asks will change, the training they receive will change and the relationships among disciplines will change to be bidirectional as patients are followed in a collaborative and integrative manner.

The data on the pervasiveness of functional symptoms is startling [2, 4, 5, 9, 15–23], as is the preponderance of functional symptoms in the primary care practice that go unrecognized or untreated [10, 18, 20]. These symptoms may lead to significant psychiatric disorders in the adult population [11, 24, 25]. In an adult primary care population, anxiety and depression are both prevalent and disabling [26]. Appropriate recognition of functional symptoms in childhood may lead to management of the early manifestations of these disorders [27] with significant reduction of suffering and financial cost down the road.

Increased awareness among PCPs of the long-term consequences of undiagnosed, misdiagnosed, and untreated functional symptoms in the pediatric population can allow for more accurate diagnosis and treatment. The facts that 20 % of the pediatric population is in need of more or different mental health care, and one in ten children report recurrent unexplained physical symptoms to their PCP [11] should be a clarion call to action for medical caregivers to both better assess and coordinate treatment.

There are numerous estimates that the need for mental health treatment greatly exceeds the availability of care [11, 12, 14, 25, 28]. This need has been reported worldwide and across cultures [18]. Creative solutions must be sought for the sake of the future health of today's youth [29].

The large-scale challenge for mental health care and preventive treatment, which is a natural outcome of early interventions, within medical primary care leads to the question of what exactly the primary provider can do to prevent long-term mental health sequelae [20].

Practical Application

The pediatrician, family physician, and other primary care clinicians usually are the first people to whom children and their families turn with health questions. They are well trained at recognizing problems of a mental health nature when the problem is severe, when parents are explicit about concerns, or when social factors such as poverty, neglect, and vulnerable age (e.g., young adolescents who become sexually active) are considered [25]. Diagnostic matters become a little more difficult when a functional symptom masks an otherwise more recognizable mental health symptoms. For example, the average PCP does not view a functional paralysis as a possible response to a psychologically based trauma, unless well versed in neuropsychiatric conversion disorders. On the other hand, the absence of evidence for a medically based symptom invites the quick relegation of a symptom to the psychological realm. The integrative shift from "it's a medical problem" to "there are psychological factors that need to be addressed" will preserve the doctor/patient relationship that is crucial in addressing especially complex symptoms.

Functional disorders are not always recognized as part of a psychosocial disorder although overall attitudes of PCPs are positive for treating psychosocial problems [10]. Sometimes, the diagnosis and treatment go beyond the scope of the PCP's practice, and a referral is preferable or even necessary [20]. Determining whether a referral will need to be made or not, can be viewed as a matrix; the alignment of various factors on the surface of Rubik's cube, which resides within a system's framework. Consideration of such a matrix can assist in the management of patients with functional symptoms. For example, the FACTR matrix (Fig. 18.1) includes F, the origin of the identification of the functional problem; A, the assessment or diagnosis of the functional symptom; C, conveyance of the diagnosis (critical to acceptance of both the diagnosis and treatment choices); T, Treatment and training options for patient and PCP; and R, Resources, referrals and maintaining follow-through relationships with both patients and other providers.

Identification of Functional Symptom

There are four possible entry points for identification of a functional problem each somewhat determined by the developmental stage of a youngster.

1. PARENT: Most prevalent is the parental (or guardian) perception of a problem [30]. Parents usually are the first to recognize symptoms and to initiate care for the child and the PCP is the first consultation. Of particular importance in this entry point is the opportunity to note any characteristics of the parents or the dynamic of parent and child that might contribute to a functional symptom. Anxiety of mothers can translate to anxiety symptoms in children. The attitudes of parents about illness (physical and mental) also may be mirrored by the children.
2. SCHOOL: Staff members at school or community-based programs may see a functional problem and alert parents; or the school

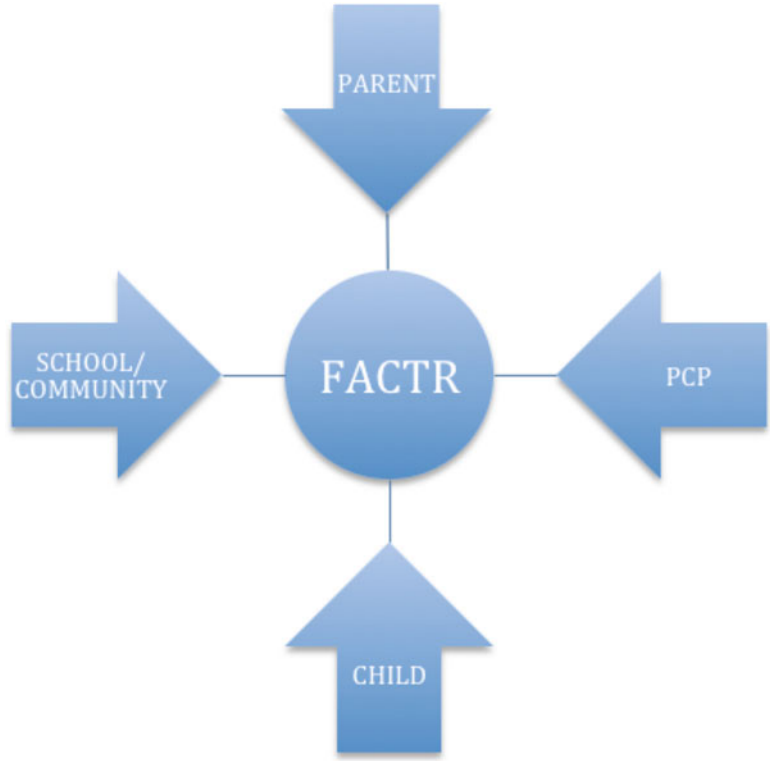

Fig. 18.1 Entry points of child/adolescent and the FACTR model for a systems approach to referral. F, Functional symptoms—Identifying the symptoms; A, Assessment—Medical evaluation, listen, interview, screen, symptom inventories; C, Conveying diagnosis—building trust, rapport, language choice, framing the referral as a collaboration; T, Treatment choice—Mental health training for the PCP, consultation; R, Referrals—Developing Resources, language of referral

may intervene after a parent identifies a functional symptom [30]. School phobias that present as somatic complaints including stomachaches and headaches are the most common functional disorders in children that may mask a mental health concern. Some school phobias mask separation anxiety and others relate to school-related traumas. An example of a community-based "diagnosis" of a functional symptom occurs when children who have extreme sleep difficulties at an overnight camp are identified with anxiety disorders.

3. PCP: The PCP may be the point of entry for identification, particularly when the PCP asks psychologically oriented questions or uses screening tools that alert him/her to a mental health-based functional symptom. A psychosocial issue may be contributing to a functional symptom, such as stomachaches that serve to keep a child from a bullying situation at school. Many PCPs welcome the opportunity to detect and address psychosocial issues [10].
4. CHILD: The child or adolescent may self-identify a functional symptom to any of the above adult resources: a parent, school, or community-based personnel or the PCP.

Assessment and Diagnosis

A comprehensive assessment [18] is the obvious starting place for all diagnoses and assures the patient that symptoms are being taken seriously. When the PCP suspects a functional symptom, it is important to hold onto this hypothesis while proceeding with the usual medical history and physical. The most recent interpretation of functional symptoms postulates an organic component when viewed from the perspective of the brain as the organ of perception. To optimize detection of functional symptoms in primary care [31], the PCP will need to look beyond physical symptoms and consider contributing psychological factors such as stress, anxiety, and depression

Table 18.1 Questions to ask when presence of a functional symptom is suspected

1. When did you start to have these (*headaches, stomachaches, tremors, other symptom*)?
2. Are you worried about these (*headaches, stomachaches, tremors, other symptom*)?
3. Can you say what the worry is? Can you put the worry into words? (Do not prompt, but leave open ended.)
4. Are others worried about these (*headaches, stomachaches, tremors, other symptom*)? How do you know they are worried? How do they show their worry?
5. What other things were going on in your life when these (*headaches, stomachaches, tremors, other symptom*) began?; Anything new or different at school?; At home?; With your friends?; In your family?; Anywhere else?
6. Have you had these (*headaches, stomachaches, tremors, other symptom*) at any other time in your life? What helped them to go away? (These questions begin to plant the seed that the symptom can be diminished or resolved.)

[25]. Additional training and tools to assess young patients [20] with complicated presentations involving functional symptoms can assist a PCP to neither miss a diagnosis nor overpathologize a symptom. It has been noted that both ignorance and biases about functional symptoms can lead to either extreme [22]. While psychological disorders may be masked or missed, sometimes the reverse occurs, and physical pathology is missed because of a presentation with overwhelming psychological overtones.

Young children are less likely to volunteer information; therefore, skilled, nonthreatening inquiry is necessary. Taking a careful history of psychosocial factors can improve the accuracy of assessment and the detection of functional symptoms [18]. Often symptom onset of a functional disorder corresponds with a noteworthy psychosocial event such as a separation, loss or death of a loved one, or even a seemingly small event, from an adult's perspective, such as a move to a different neighborhood or new school. Questions to guide the PCP through an assessment of psychosocial factors are listed in Table 18.1.

Psychological screening tools [18] can be useful to alert a PCP to contributing emotions, but they cannot replace the relationship between PCP and patient. Used judiciously, a simple screening tool, especially with adolescents can alert the PCP to depression and anxiety, social difficulties, and suicidal thoughts. A review of the literature of 95 mental health screening and assessment tools conducted by UC Davis and compiled by Shannon Williams [32] recommended three tools that are brief, have a wide age range and that PCPs will find helpful. These are listed in Table 18.2 along with those that are used by physicians as part of their interviews of children and adolescents. It is notable that Vallance [25] reports studies in the USA and Australia showing that many young people have contact with their PCP in the month before a suicide or self-harm; however, fewer than 50 % of all PCPs in the study routinely screened for suicide risk.

When gathering a history and asking questions in order to determine if a symptom is functional, an empathic and nonjudgmental manner yields favorable results. Children need to feel heard, to have their symptoms validated, regardless of their origin, and to be given some sense of partnership in the journey of discovering what is making him/her feel sick. It is beneficial for children as well as their parents to be educated about the complexity of the mind. They can be taught that the brain can alert us through the somatic symptoms that something needs attention and may need to be fixed, even if the problem is a thought or worry. In the adult population, cognitive behavioral therapy (CBT, Chap. 19) has been effective in treatment of functional somatic symptoms [7]. The PCP, with similar success, can offer simple corrections of distorted ideas about illness with children. Scripted dialogue that can guide sensitive psychological questioning may be useful for the PCP [20]. For example, after listening to a youngster describe his presenting problem, the PCP might say: "You have done a very good job describing what has been going on, and I wonder if you can tell me when this started? Was there any significant event that was happening at that same time?" Or when getting ready to make a referral, the PCP might say: "I am glad you are comfortable sharing this information with me and that you recognize it needs to be addressed. I know just the person that will be able to help you with what you are experiencing, while we complete the studies here."

Table 18.2 Traditional screening tools options

Name of tool	Authors	Description	Format	URL
BESS—Behavioral and emotional screening system	R.W. Kamphaus, C.R. Reynolds	Measures strengths and weaknesses, 3–18 years, Preschool-Grade 12, reporters are parents, teachers and youth The BASC-2 Behavioral and Emotional Screening System (BESS) Student Form is a recently developed youth self-report rating scale designed to identify students at risk for behavioral and emotional problems. The BESS Student Form was derived from the Behavior Assessment System for Children-Second Edition Self-Report of Personality (BASC-2 SRP) using principal component analytic procedures and theoretical considerations	Teacher form Self-report (grades 3–12) Parent form	Must purchase: http://www.pearsonassessments.com/HAIWEB/Cultures/en-us/Productdetail.htm?Pid=PAaBASC2bess
PIC-2/PIY/SBS-personality inventory for children, 2nd edition	D. Lachar, C.P. Gruber	Student Behavior survey—ages 5–19 years; screens for mental health, behavior problems, and academic/school issues This highly regarded test is widely used by clinical and school psychologists to evaluate the emotional, behavioral, cognitive, and interpersonal adjustment of children and teens. The PIC-2 is one of three coordinated instruments: It provides the parent's description of the child; the PIY provides the child's self-report; and the SBS supplies a teacher rating. While each of these instruments has been validated to function independently, together they provide an integrated picture of the child's adjustment at home, at school, and in the community	Parent form	http://portal.wpspublish.com/portal/page?_pageid=53,112601&_dad=portal&_schema=PORTAL
Personality inventory for youth (PIY)	D. Lachar, C.P. Gruber	9–19 years; screens for mental health, Behavior Problems, academic/school issues In the tradition of the highly regarded *Personality Inventory for Children, Second Edition* (PIC-2), this self-report measure assesses psychological problems in 4th–12th-graders. The *Personality Inventory for Youth* (PIY) answers the need for a multidimensional, psychometrically sound self-report instrument designed specifically for young people.	Self report	http://portal.wpspublish.com/portal/page?_pageid=53,102890&_dad=portal&_schema=PORTAL
CAPS-the child/adolescent psychiatry screen [28]	J.Q. Bostic	Ages 3–21, 15–20 min, parents complete-target symptoms and disorders	Parent form	http://www2.massgeneral.org/schoolpsychiatry/ChildAdolescentPscychiatryScreenCAPS.pdf
The pediatric symptom checklist	M. Jellinek, J.M. Murphy, S.J. Bishop, M. Pagano	Ages 6–16 A brief screening questionnaire used by pediatricians and other health professionals to improve the recognition and treatment of psychosocial problems in children	Parent form	http://psc.partners.org/psc_english.PDF

Name of tool	Authors	Description	Format	URL
Child behavior checklist	T. Achenback, L. Rescorla	The CBCL, used for evaluating children and adolescents ages 6–18, measures problems with aggressive behavior, anxiety/depression, attention, rule-breaking behavior, social interaction, physical complaints, disordered thought, and withdrawn/depressed behavior. It is used for initial assessment and can also measure changes in behavior over time or following a treatment.	Parent form Teacher form Youth self-report	Must purchase: http://www.aseba.org/
		The Parent Checklist is one of the most widely used parental ratings for behavioral problems and social skills in children. For evaluating children younger than age 6, the Child Behavior Checklist/1½–5 is used instead. The CBCL/1½–5, preschool form, obtains parents' ratings of 99 problem items plus descriptions of problems, disabilities, major concerns about their child, and the child's strengths. It also includes the Language Development Survey (LDS) for identifying language delays.	Clinician form Preschool form	
		Versions for parents, teachers, and youth each contain approximately 120 items and take 15–20 min to complete. The clinician form (DOF) contains 96 items and is for children and adolescents ages 5–14		
GAPS questionnaires		Ages 11–21	Younger adolescent form	http://www.ama-assn.org/ama/pub/physician-resources/public-health/promoting-healthy-lifestyles/adolescent-health/guidelines-adolescent-preventive-services.page
		Screen for adolescent psychosocial and behavioral issues	Middle/older adolescent form	
			Parent/guardian form	
The HEADDS psychosocial interview for adolescents	J.M. Goldenring, E. Cohen	Provides an outline for conducting an adolescent psychosocial interview	Interview outline	http://www.bcchildrens.ca/NR/rdonlyres/6E51B8A4-8B88-4D4F-A7D9-13CB9F46E1D6/11051/headss20assessment20guide1.pdf
Mental health screening and assessment tools for children [32]	S.T. Williams	This resource looks at 95 different tools and assesses each in terms of use, population, age, time, and reliability		https://docs.google.com/viewer?a=v&q=cache:KsqDf8LAotIJ:humanservices.ucdavis.edu/academy/pdf/final2mentalhealthlitreview.pdf+&hl=en&gl=us&pid=bl&srcid=ADGEEShYY2-CuDpGjWC8EtUW2kiZn8RYKk-OctRBr-PLRPO2Q3uYsiskr2cnVTgNrI_O6OHXMXFAbX_r8ylFNrtT6rczCKqKOTB7qSKsiBhGn6uPQtwK8qrO_aOGXGHGtsW-lbiwDKl3E&sig=AHIEtbTIijKp_aKvxqRBSkaVsxY_W_bzjw
Vermont guidelines		This resource contains the most common screening tools used by pediatricians		http://www.google.com/url?sa=t&rct=j&q=&esrc=s&source=web&cd=2&ved=0CDYQFjAB&url=http%3A%2F%2Fhealthvermont.gov%2Ffamily%2Ftoolkit%2Ftools%255CI-6%2520Assessing%2520Emotional%2520and%2520Behavioral%2520Health.pdf&ei=d16tULvvCrDG0AHCsoDgBA&usg=AFQjCNEKP3vrljTRGtFUDqMtk7Mb4v9jfg

Conveyance of Findings

The patient and family need to be educated about the PCP's impression and working diagnosis when functional symptoms are being evaluated. The constructive use of language is paramount in order to educate the patient and the family about relevant medical and psychological findings, to strengthen collaboration, and to ultimately enhance treatment compliance. Our word choices are a powerful mediator, especially in the medical realm, and can increase or decrease the therapeutic relationship [2, 3]. For example, consider the difference between a "poke" and a "shot" or a "needle." Even more unintentionally hurtful is the statement "You will just have to live with that pain," which the child may hear as, "You will die without the pain." In order to facilitate treatment and maintain collaborative relationships with patients, it is important to use care in language choices to describe and discuss somatic symptoms [5]. The way in which this information is imparted to patients and their families has failed at times to facilitate treatment [33, 34]. This is one of the reasons for the change from such pejorative words such as hysterical or conversion symptoms to the current more neutral and educational term, "functional" [2].

There are suggestions inherent in all communication and these intended (and sometimes unintended) suggestions may either inhibit or facilitate a positive response to a mental health referral. For example, saying, "Dr. S is an expert with this type of situation, and I am confident she will be of help to you," is a very positive suggestion that communicates hope. Alternatively, "Dr. S. may be able to help you with this problem" communicates doubt about the potential effectiveness of a referral. Careful wording to foster positive responses is warranted.

The question of what terminology we use is one of many factors in encouraging parents and their children to access mental health services as needed. Parents are likely to repeat the PCP's words when discussing the nature and meaning of the symptoms with their children. They are also likely to convey their own beliefs about the symptoms. In one family that practiced a fundamentalist approach to their religion, the child's symptoms were seen as the work of the devil; in another the symptoms were seen as punishment for the child's misbehavior. Therefore, it is useful to coach parents on what to say to their children. Working with pediatric patients requires attention to the dynamics of the family [35]. If there is moderate to severe family pathology, such as discord, irrational behaviors, lack of emotional resources, or other signs of inappropriate parenting, referral may be even more pressing and at the same time less likely to be accepted.

Treatment Planning

When it is time to decide how best to proceed with treatment, the PCP has several options: to obtain consultations regarding how to treat, to build a repertoire of psychological treatment skills of their own through professional education, and/or to refer for mental health treatment. There is a shortage of affordable mental health care. To ameliorate this problem many excellent suggestions have been made [25] including provision of more in-depth training of PCPs in mental health diagnosis and treatment to facilitate their assuming a role of treatment. Instruments can be provided for diagnosis and assessment of symptom severity. Frequent and timely consultation with mental health experts is key [26, 28] as is making changes in the current care model to emphasize collaborative care.

The PCP who builds a set of mental health skills will be better at diagnosing and assessing functional disorders and able to treat many functional presentations. Through use of brief interventions such as CBT to address erroneous beliefs, enlisting parent support, learning medication management of mental health symptoms, and acquiring a repertoire of stress reducing techniques, the PCP can address many functional presentations in the pediatric population. This requires that the PCP is willing to explore emotional issues with the patient, and some are reluctant to do so [25] due to lack of skill, experience, or time. In these instances, the PCP can develop a network of

resources [20] that can provide consultations, coach with psychological treatment skills, or serve as referral sources.

Relationships, Resources, Referrals

What are the resources available to the PCP? And when using those outside resources what happens to the role of the PCP in continuity of care? Seeking out mental healthcare providers who are available to consult about treatment needs and options requires time and effort in building relationships, which is an investment with large returns. Sometimes, no local resources are available, and this may be a major motivator for the PCP to learn more skills or find human resources who will consult/coach from a distance. The patient or family's perception of the stigma of mental illness [18] may impede referral and motivate the PCP to manage the patient in the primary care setting.

Current models for psychiatric or psychological treatment are largely built on single consultant-single patient office visits, with cost and availability both contributing factors that limit access to care. PCPs may refer to psychiatrists, psychologists, social workers, and other counseling resources but the impediment of the cost of mental health treatment has not been adequately addressed in the research [18]. While it is cost saving for the PCP to provide treatment, often the parents and/or child fail to recognize the PCP as a resource for emotional difficulties [25]. However, consultation for the PCP is less costly than referral [25] and can be a next step.

The Massachusetts Child Psychiatry Access Project (MCPAP) [28] broke new ground when it tested a model that could change primary care in pediatrics. In this well-designed project, psychiatrists were available for consultation on difficult cases when functional symptoms (1) needed to be assessed; (2) needed to be differentiated from other medical or mental health symptoms; (3) could be addressed with simple or brief treatment interventions by the PCP; or (4) required referral for further more intensive treatment. The PCPs who reported they could usually meet the needs of psychiatric patients increased from 8 to 63 % after utilizing the consultation service for help with diagnosis, referral sources, and medication choices. Although significant barriers to PCP detection of mental health problems in children remained, such as limitation of training, time, and specialist availability, the MCPAP project concluded that a collaborative model between PCP and Psychiatry would augment detection and treatment.

As integrative care models are developed, the PCP's taking the role as "case manager" has shown promise for the best follow-through [28, 36]. The PCP can function as the hub and liaison in continuity of care as he or she is likely to have known the child the longest and thus is able to spot patterns of behavior both in the child and in the family that can guide diagnosis and the flow of treatment between medical and mental healthcare providers.

Regarding resources, the success of referring a patient who presents with functional symptoms depends on the PCP's consideration of (1) availability of mental health care, (2) cost of effective treatment, (3) education of the physician to parse out those who will benefit from a referral [20] from those that can be treated by the PCP, and (4) education of the patient regarding treatment interventions.

When Are Referrals Most Likely to Be Needed?

When the problems do not improve with initial interventions by the PCP or the child has more significant impairment or coexisting conditions then specialty treatment is required [36].

Ellen [18] suggests referral if symptoms fall in one of three categories:

1. Severity of symptoms: If there is functional impairment such as found with severe ADHD, severe depression, or anxiety.
2. Diagnostic difficulty: If the PCP is unsure of diagnosis.
3. Responsivity to treatment: If there is a failure to respond to treatment or nonacceptance of recommended treatment.

What Is Needed for the Referral to Be Successful?

Once the decision to refer is made, then the PCP must consider how to manage the referral. If the PCP already considers mental health to be part of the overall health of a child, this will be more familiar. If the PCP is uncomfortable with addressing mental health issues, then moving the mental health referral from the "end of the road" paradigm to part of much more productive matrix, requires attention to many factors.

Some integrated practices bring mental health services into the primary care setting [18], either as part of the team or on an "on call" basis, and build referral resources. These relationships are likely to improve the chance of success for patient referral.

The quality of the relationship between physician and patient is associated with adherence to treatment, parent satisfaction with care, and sensitive discussion of psychosocial concerns [20]. When recommending referral, the PCP who uses open empathic communication is compassionate and provides psychoeducation about the emotional factors of all illness is likely to be effective. The PCP can discover the patient's beliefs and fears about mental health services and demystify the nature of mental health [18]. Culture, race, and ethnicity may be factors that affect how parents receive mental health advice from a PCP [37]. For example, in some cultures there is a belief that the family takes care of all problems and it is a matter of shame to seek counseling outside the family. In this instance the PCP might refer the entire family and suggest that they are already good at solving problems as a unit and might consult with an expert who could then assist the PCP to better address the problem. A social systems framework applies general systems theory to social settings. It identifies the structure, interactions, and goals of those social interactions. This attention to social context will permit the PCP to include such considerations when making a referral. Familiarity with one's community, its diverse customs and beliefs, as well as the resources within the community can facilitate the referral process.

Case Studies

These case studies will be presented from the perspective of the treating mental healthcare provider (a psychologist) with special consideration given to the intricacies of decision-making by all participants in the referral process. It is recommended that the reader consider at each phase of the case how either side of the referring process made its decisions.

Case #1

A 5-year-old boy was brought for treatment to the mental healthcare provider "as a last resort." In the telephone intake, the mother stated her son had an "acute onset of migraines" during a family vacation hiking at high altitudes. She said she had been to the PCP and several neurologists (including a pediatric neurologist) over the previous 6 weeks. She stated that her child experienced constant pain the entire time. The mother sounded desperate for help. Some medication had been given with little result, but the most recent neurologist had told mother and child, "You will just have to live with this." One of the physicians had suggested consulting either a psychiatrist or psychologist, and she had networked with friends to find one.

A full psychosocial history was gathered from the mother and treatment options were discussed. She was extremely anxious about her son's condition. The therapist explained relaxation techniques and hypnotic pain management strategies to her and said he would teach her son these techniques. The therapist further explained that physical pain is real even when no clear causative factor can be determined and that her son could learn skills to feel much better.

On first meeting the young boy he entered the office, with his head turned down toward the floor, and when the therapist said, "Hello, how are you?" he responded, "I'm an 8." The therapist quickly realized that the boy was expecting yet another physician to examine him and was giving a pain measure report. He responded with, "Well,

would you like to make that a 4?" He looked up at the therapist and said, "How about negative 100, or negative 1,000, or a trillion, or eternity?" His response indicated several important elements. He was now interested in what we could be done to change his situation. The therapist had captured his attention with something hopeful. Also, he was ready to make the pain go away, cutting it in half was not enough. And finally, whatever eternity meant to him it seemed an odd comment in this context and suggested there might be a contributing recent event having to do with life and death that might be troubling him. "OK," replied the therapist. I think we can do that together."

The therapist then quickly taught him a hypnotic induction in which he put the discomfort in his head inside an imaginary balloon and released the balloon into the air. He did some psychoeducation about headaches and migraines, assured him that the body knows how to take good care of us, and elicited from him several personal experiences that proved this to him. (I fall down, get hurt, and then I heal).

The boy left the office with a bounce in his step. His mother called that evening to ask what the therapist had done. "This is the first time in 6 weeks that he has no pain," she reported.

Questions

1. When pain resolves this quickly, should we consider this a functional somatic symptom?
   (a) No. Functional somatic symptoms take time to resolve.
   (b) There is no adequate research to answer this question.
   (c) A typical functional somatic symptom lasts 3–6 months in children.
   (d) This kind of functional somatic pain can resolve quickly with hypnosis.
   (e) Both (b) and (d).
2. What feedback might the mental healthcare provider give to the PCP?
   (a) Information on the mother's anxiety.
   (b) Resources for yoga, acupuncture, or other stress reducing skills.
   (c) A written report with treatment outcome.
   (d) All of the above.
   (e) No feedback is required.
3. What else might the PCP have done with this 5-year-old prior to a neurological referral?
   (a) Ask about contributing psychosocial events
   (b) Ask the child what would be different if the pain had never happened.
   (c) Educate the child about ways the mind and body work together.
   (d) Ask the child what advice he might give to a friend with his problem.
   (e) All of the above.

Answers

1. (e): The definitions of functional somatic pain are evolving. Research on the time element is sparse, but cases of quick resolution with hypnosis are reported in the irritable bowel syndrome (IBS) and migraine literature. The PCP will benefit from familiarity with this research and could consider training in pediatric hypnosis. Identification of mental healthcare providers who are trained in hypnosis is an important resource to develop.
2. (d): A written report from the mental healthcare provider would facilitate communication and care for the child. Alerting the primary care provider about the mother's anxiety can be useful since children are reactive to their parent's anxiety and can absorb those feelings. The mental healthcare provider can suggest that the PCP encourage the parent and child to learn techniques to reduce anxiety and provide some resources to do so.
3. (e): The way in which the PCP speaks to a child about the pain problem can hinder or facilitate the symptoms and the referral process. The PCP wants to communicate that this is a mind and body interaction, many things can cause us somatic pain, and that pain can be scary and add to our discomfort. In addition, eliciting ideas from the child signals that there are solutions and provides hope, which is a necessary ingredient to healing.

Case #2

A 10-year-old girl was referred for psychological treatment by her pediatrician because of ongoing stomach aches that appeared to be of a functional nature. The pediatrician had a long history of referring patients to the psychologist. Interviews with each of the divorced parents painted a picture of a very quiet, shy, smart youngster who had only a few friends. She complained of stomachaches most school days and asked to stay home. The parents rarely allowed her to miss school. Teachers reported her as quiet and retiring in the classroom. She had little psychological awareness and neither did her parents although they considered her stomachaches to be stress related. The psychologist had verbal contact (via telephone) with the school personnel and parents but not with the pediatrician.

Treatment focused on teaching stress reduction techniques, body awareness, and fostering confidence through learning new behaviors in interpersonal relating. Treatments included a mix of operant conditioning for building confidence (learning to raise her hand to answer classroom questions), CBT, self-hypnosis, and supportive psychotherapy. Within about 3 months of the commencement of treatment the complaint of stomachaches reduced significantly. By 6 months there were no more complaints. Several major life events occurred, including mother's remarriage and the birth of a half-sibling and the patient remained symptom free during these seemingly happy life stresses. Treatment was reduced to once a month through to the successful start of the next school year at which time the patient was discharged with instructions to parents to call if the symptom resumed. The parents communicated the results of treatment to the pediatrician.

This case represents a fairly common mental health presentation in the pediatric population. The presence of somatic symptoms without major medical etiology is typically thought to be related to some distress in the child's life expressed through muscle tension and the "second brain" as the gut is sometimes called. How many of these untreated cases go on to develop IBS is an interesting question worthy of more research as there has been a proliferation of knowledge in the successful treatment of IBS with hypnotic suggestion.

Questions

1. What are the factors that needed to be considered in order to refer?
   (a) Medically unexplained symptoms, parental and teacher concerns.
   (b) Concerns about future development of IBS.
   (c) Family openness to referral.
   (d) Availability of mental health resources.
   (e) All of the above.
2. What else might the pediatrician have done to assist the child to address the symptoms?
   (a) Do a psychosocial evaluation with the child and parents.
   (b) Assess anxiety and depression and reassure the child.
   (c) Teach the child relaxation and stress reduction techniques.
   (d) Explore school-related and community resources for stress reduction.
   (e) All of the above
3. What kind of follow-up might the pediatrician do with the patient?
   (a) Meet with the child.
   (b) Call the parents.
   (c) Speak with the mental healthcare provider.
   (d) Talk with the schoolteacher.
   (e) No follow-up is required.

Answers

1. (e): This was a good case for the pediatrician to refer. There were no clear medical reasons for the symptoms. He knew the family and trusted the parents' suspicions and teacher concerns that the symptoms were related to some emotional issue. He had inquired about the timing and nature of the symptoms and knew that early morning tummy aches on school days were often related to school anxieties. He was concerned that untreated these symptoms could lead to chronic IBS. He had a relationship with the mental healthcare

provider and knew he could expect feedback if needed. He was aware the family would be open to a referral as they were asking for help and thought the stomachaches had an emotional basis.

2. (e): The primary care provider has many options to pursue once it is clear that a symptom is functional. Not all of these might be needed, but any could be helpful. The child had multiple psychosocial issues occurring at home and if the school counselor could offer a program for children of divorce, as many do, this might have been a resource. Empowering a youngster with skills to calm and sooth builds confidence and character strength. The trained PCP can teach these skills and often a referral to a mental healthcare provider will be unnecessary.
3. (a): The primary care provider as the ongoing case manager would meet with the child during a brief follow-up appointment to discuss how the referral is progressing and to assess the child's improvement (or not) and provide through this behavior the message that the patient's progress is important and will be monitored.

Case #3

The following case is included as an example of how complex diagnosis and treatment can be for both PCP and the referral source and to exemplify opportunities for enhancement of collaboration and patient–doctor relationship in integrative models of care.

A 15-year-old female presented to her PCP with an acute onset of migraines. She was so debilitated by pain that she was unable to attend school. She complained of constant pain, inability to sleep, nausea, and vomiting. She was unresponsive to prescribed medication. The pediatrician referred her to a neurologist for assessment and did not follow up with the young adolescent. The neurological findings reported acute migraines, and the neurologist suggested mental health intervention to teach pain management and assist in the return to normal activities such as school. The referring neurologist chose to address the dysfunctional behavior, not going to school, and had nicely couched this in terms of pain management.

At the time of referral, the adolescent was being tutored at home each afternoon. The family found the mental healthcare professional by themselves. The psychologist requested a copy of the neurology report to review as part of the evaluation and assessment.

The psychologist's interview with the parents prior to meeting the youth, suggested very worried parents in search of help for their daughter. There was a history of migraines in the family as well as previous migraines in the daughter, in contrast to the acute onset that had been reported. The parents could not identify any precipitating events that might explain the migraines as part of a stress response, but they did note as a curiosity that the migraines over the past 2 years had not been present during vacation or family travel.

The interview with the 15-year-old revealed a youngster who did not appear to be in pain in spite of her verbal report of an 8 on the 10-point pain scale. None of the typical signs of distress or discomfort were noted, such as fatigue, irritability, facial distress or tension, or motor retardation. Further, she did not seem upset by the inability to attend school or see her classmates. This was particularly important since she was described as a bright, high achiever and the school threatened she would need to repeat the entire school year if she did not attend or be tested in her subjects. She refused to take any tests and said she could not read because of double vision. It was noted that her behavior seemed more like a younger adolescent's and that there was enmeshment between mother and daughter. The "La Belle Indifference" or what might better be described as a mismatch between reported symptoms and displayed psychological features suggested a working diagnosis of a conversion disorder, with possible school phobia. Puzzling about her presentation, however, was her unusual amount of empathy and emotional intelligence about both her family and friends. Most conversion disorders are characterized by a lack of insight.

Treatment proceeded in concert with school officials and with the treating psychologist

managing the case. There was no contact with the PCP or neurologist nor had they requested feedback or collaboration. The psychologist instructed the parents to enroll their daughter in the after school short-term academic program the school system provided. She also recommended a psychiatric consultation to consider medication that would address the anxiety apparent in the test taking and school phobic behaviors. Medication was begun with an emphasis on its usefulness in managing pain quickly. Family therapy was recommended and started. Twice weekly intensive psychological treatment included teaching stress reduction techniques, exploring intra and interpersonal dynamics with no mention of the migraines or double vision as attention to conversion disorder symptoms is usually unproductive.

The young lady had a rough start back to the school-based program. In her first 2 weeks she attended only 3 days. She reported anxiety attacks before getting on the school bus, with vomiting and dysphoria. The parents were encouraged to ensure that their daughter get on the bus no matter what, and the school offered to send an aid to the home to assist with this. The parents declined the offer of extra support, but the prospect of outside school involvement strengthened their motivation and resolve to get their daughter on the bus to school. It is highly recommended to follow a team approach, which can successfully address the many factors of a biopsychosocial framework in complex functional disorders.

Several events were uncovered during treatment that quickly resolved the adolescent's symptoms. After being caught in the act of stealing from the treating therapist, the behavior was explored in a safe and gentle manner. This allowed the girl to reveal she did not have migraines, never had, and had simply adopted that symptom to get out of school. She was appalled at her own willingness to have submitted to some invasive testing even though she had no real physical symptoms. She described herself not as school phobic, although she did feel anxious and perfectionistic, but rather afraid to face her peers because she had such a long history of telling lies that she was often caught in a web of lies she could not support. The course of treatment was reformulated to teach assertiveness and support honest behaviors as well as building resources for confidence and resilience. The young woman, of her own volition, asked to attend summer school, to take the necessary tests, and to get more help with skills to reduce her anxiety and mild OCD.

This unusual case highlights several vulnerabilities in the assessment of functional symptoms. In the absence of clear physical or biological evidence or markers of illness, the physician has only the symptom report of the patient. While the patient improved markedly, examination of the case suggests many junctures where communication can falter and the patient may not receive beneficial treatment.

Questions

1. What are the possible reasons the neurologist suggested mental healthcare referral?
   (a) The neurologist did not see improvement in the report of pain.
   (b) The child was missing a lot of school.
   (c) The child did not seem to mind being out of school.
   (d) The neurologist wanted the child to learn pain management skills.
   (e) All of the above
2. What assessment instruments might have been used by the referring neurologist to find or rule out concomitant mental health issues?
   (a) A clinical interview with the patient inquiring about recent stressful events.
   (b) A brief scale that measures anxiety, depression, and/or traumas.
   (c) A clinical interview with the parents and patient and use of assessment scales.
   (d) An assessment of hypnotizability or imaginative involvement.
   (e) None of the above.
3. What role could the primary pediatrician play in the management of this case?
   (a) Collecting the reports from the neurologist and mental healthcare provider
   (b) Phone or written contact with the mental healthcare provider.

(c) Supporting the neurology referral for mental health care.
(d) Follow-up contact with the child.
(e) All of the above.

Answers

1. (e): Neurology is an area of medicine that sees a lot of functional symptoms. Since the child was not responding to medication, suggesting an alternative path to reducing discomfort provided an acceptable and non-stigmatizing entrée into the discussion of how a mental healthcare provider could help. Using the goal of returning to "normal" school routine was appealing to the parents, especially with the "La belle indifference" toward school that the child exhibited. While the neurologist might have taught the child pain reduction techniques, there was a suspicion of other emotional underpinnings that needed to be explored, beyond what the medical interview yielded.
2. (c): Experienced neurologists inquire carefully about the timing and nature of recent events when there is an acute onset of severe medically unexplained symptoms. They do this with the child as well as with the parents using a family system model. Adding a developmentally appropriate standardized assessment tool can be helpful for patients who are willing to fill them out. Increasingly, hypnosis has been used as a tool to explore subconscious contributions to a symptom. This is an extremely valuable tool to employ with any functional symptom to aid in its diagnosis and treatment. A neurologist who uses hypnosis also may use this as a way to introduce some pain management suggestions. This may be done by either a hypnosis-trained PCP or mental healthcare provider.
3. (e): When the primary care provider has an ongoing good relationship with the patient, he or she is best positioned to oversee the implementation of the entire treatment plan. Communication includes seeking reports and information from the other treating parties and contact with the other treating parties to facilitate the primary care of the patient. When the primary care provider has asked for the consultation with neurology, it is important to support the referral the neurologist suggests. This provides a sense of team work to the patient and family and provides reassurance that each is doing what is necessary. It also provides the implicit framework that the symptom is both mind and body. The treating psychologist can also facilitate communication among providers by requesting reports and contact as well. When professionals at either end of the medical-mental health continuum reach out to one another about a case they have in common, the patient and his or her family can benefit.

Additional Training

As noted in the FACTR model, a children's care provider can do many things to improve the outcome of their child and adolescent patients who present with functional symptoms. It can be very helpful to receive training in simple and quick tools for screening of emotional disorders and to develop a plan of action for implementing help or referral as needed.

Specific short courses in behavior modification, CBT, hypnosis, and integrative medical techniques will teach the PCP new interventions with children and their families. The PCP then can, in turn, teach parents behavior modification and counseling strategies to improve their parenting skills.

The remainder of this book has detailed information on a variety of techniques that can be employed in clinical practice by the PCP before considering a referral.

Conclusions

The PCP who treats children with functional somatic symptoms must assess, diagnose, treat, and sometimes refer those young patients. The FACTR model provides an acronym for the factors that enhance treatment outcomes. The

nature and terminology of functional symptoms is evolving. Current models attempt to include the systemic psychosocial factors involved in functional symptoms and reverse historical trends of mind/body dualism that confuse patients and stigmatize mental health. Patients benefit when the PCP hones his or her skills to assess the psychological contributions to illness and then to convey diagnosis in a compassionate and empathic manner. Acquiring a variety of mental health treatment skills builds the PCP's treatment repertoire and enables improved identification of patients who require referral. Referral sources work best when, as with patients, relationships are nurtured. This may take the form of meeting with the referral sources face-to-face, participating in phone calls to discuss a case, or requesting follow-up letters from the referral source. The collaborative model benefits the child and family. The PCP can remain in the primary role for continuity of care, supporting all involved, and especially the child and his or her family. This model is especially important for referrals of children with functional somatic symptoms that appear "medical" and yet may be better understood as physical manifestations of psychological forces. Collaborative and integrated healthcare models in which the primary care provider is in the role of case manager can be applied with much success to the treatment of functional symptoms.

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Cognitive-Behavioral Therapy for Youth with Functional Somatic and Internalizing Symptoms

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Abstract

Functional somatic symptoms (FSS) are prevalent, highly comorbid with internalizing symptoms (e.g., anxiety and depression), and cause significant impairment to youth and their families. Children and adolescents with FSS typically pursue repeated medical consultations without relief, and internalizing symptoms are rarely detected. Research has shown that cognitive-behavioral therapy (CBT), an effective intervention for internalizing disorders, is a promising alternative treatment approach for youth with FSS. Physicians can play a critical role in facilitating patients' access to CBT by identifying internalizing symptoms and promoting families' engagement in a nonmedical intervention. This chapter provides guidelines to help physicians identify internalizing symptoms and provide psychoeducation about the biopsychosocial model. A description of brief cognitive-behavioral strategies that can be implemented by physicians and their staff, as well as more specialized techniques that should be implemented by a professional with training in CBT, will be presented. Finally, two case studies will illustrate the application of CBT for youth with FSS and internalizing symptoms.

Keywords

Functional somatic symptoms • Anxiety • Depression • Cognitive-behavioral therapy • Children and adolescents • Medical setting

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Functional somatic symptoms (FSS), or physical symptoms of unknown pathology, affect 10–30 % of children and adolescents in the USA [1] and account for 1–4 % of all pediatric visits [2, 3]. The physical discomfort caused by these symptoms is commonly accompanied by internalizing disorders such as anxiety or depression [4], which are less observable than externalizing disorders like disruptive behavior disorders. Research conducted in pediatric gastroenterology indicates that about 50–80 % of children with functional abdominal pain meet criteria for an anxiety disorder [5–7]. Similarly, in a recent trial of 180 youngsters presenting to pediatric cardiologists with functional chest pain, 70 % met criteria for an anxiety disorder [8]. In addition to this data collected from clinical samples, a large body of evidence suggests that youth with FSS in community samples are significantly more likely than unaffected peers to experience anxiety and depressive symptoms [1, 9–15].

The prevalence of FSS, paired with high rates of co-occurring internalizing symptoms, is particularly concerning given the considerable impairment associated with these conditions, including poor school attendance and performance, interpersonal and social difficulties, and family disruption [10, 16–18]. In a recent trial of 40 children presenting to pediatricians or pediatric specialty clinics with FSS and anxiety disorders, 53 % missed school due to these symptoms, with an average of ten school absences over the past year [19]. This study also highlighted the chronic nature of FSS and comorbid internalizing disorders, with 70 % of youth reporting that their physical symptoms had been present for 1–2 years, and 43 % reporting duration of more than 2 years. As symptoms persist, youth tend to seek repeated medical consultations and undergo excessive, invasive, and expensive diagnostic procedures [11, 20, 21]. Throughout this process, internalizing symptoms are rarely assessed or detected [22–24], and children may suffer for years without relief. Lengthy delays between symptom onset and accessing effective treatment are alarming because childhood and adolescent FSS predicts a variety of problems in adulthood, including anxiety and depressive symptoms [25, 26], functional somatic symptoms [12, 27], and increased healthcare utilization [28].

The persistent course, high medical utilization by affected patients, and under-recognition of comorbid internalizing disorders associated with FSS underscore the need for providing effective treatment for this population. Standard care for functional abdominal pain, for example, consists of ruling out organic disease, providing reassurance about the absence of disease, and recommending dietary changes or pharmacotherapy [22]. This approach has been largely ineffective [29–32] and has driven researchers to investigate alternative approaches. Given the documented link between FSS and internalizing symptoms, psychological intervention has emerged as a promising treatment for these youth.

Cognitive-behavioral therapy (CBT) has been documented as an efficacious treatment for anxiety and depression in children and adolescents [33–36]. The literature has also supported the use of cognitive-behavioral strategies (e.g., relaxation, distraction, and contingency management) for reducing the severity and frequency of headache and abdominal pain [37–41]. The effect of CBT on both physical and psychological outcomes for youth with comorbid FSS and internalizing disorders has only recently been examined. A recent study compared participants with FSS and anxiety disorders receiving a CBT protocol emphasizing physical–emotional symptom associations [42] to a waitlist control group. Results showed that CBT participants exhibited decreased clinician-rated anxiety, as well as parent- and self-reported reductions in pain [19]. While further research is necessary, preliminary evidence supports the use of CBT as a viable alternative approach to standard care for youth with FSS and internalizing disorders.

This chapter will provide recommendations to facilitate identification and appropriate management of comorbid internalizing symptoms in youth with FSS in order to improve care for this population. Guidelines to help physicians identify and normalize internalizing symptoms in a manner that promotes acceptance of cognitive-behavioral techniques will be provided. The chapter will then explain several cognitive-behavioral strategies,

including psychoeducation, symptom monitoring, relaxation, cognitive restructuring, and exposure. While some of these strategies may be implemented by physicians and their support staff (e.g., nurses, social workers), some patients will require more time and resources than may be available in busy pediatric settings. Given the stigma that is often associated with seeking mental health treatment, we also provide tips for increasing families' acceptance of referrals to CBT specialists. Finally, CBT techniques are illustrated through two case studies, and scripts and handouts are provided in the appendices to aid in the practical application of these strategies.

Practical Applications

Identifying Internalizing Symptoms

Despite high rates of co-occurring FSS and internalizing symptoms, anxiety and depression are often undetected. Unlike disruptive behavior disorders, internalizing disorders often go unnoticed by teachers and parents because these children are generally compliant, follow rules, and do not draw attention. Relying on families to raise psychosocial concerns may be problematic since many families may not conceptualize emotional symptoms as connected to distressing physical complaints. Furthermore, research has shown that most parents do not discuss mental health issues with physicians, perhaps due to the time constraints of the visit or financial and insurance concerns for procuring mental health services [43–45]. These studies also show that parents are most likely to consult with physicians about problems that are disruptive (e.g., habits, behavior problems, family conflict). Therefore, physicians can play a critical role in facilitating effective treatment for youth with FSS by assessing for anxious and depressive symptoms.

Internalizing symptoms can be identified in a few ways. The first is to integrate brief self- or parent-report screeners of anxiety and depression into standard intake packets that can be completed while the family is in the waiting room (e.g., *Multidimensional Anxiety Scale for Children-ten item* [46], *Children's Depression Inventory*) [47], both available for order from Multi-Health Systems Inc. (sample items include *"I try to stay near my mom and dad," " I'm afraid that other kids will make fun of me," "I am sad all the time," "I can never be as good as other kids"*). These validated measures yield summary scores indicating whether the patients' responses signify clinically elevated symptoms compared to a normative sample. Alternatively, skimming responses can typically provide adequate information to determine whether further evaluation is necessary. Indications that the child is clingy with caregivers, shy, avoids difficult situations, or worries a lot may indicate symptoms of anxiety. Positive responses to questions about feeling down, loss of pleasure, or being hard on him or herself may be symptoms of a depressive disorder.

Another option is assessing anxiety and depressive symptoms during the medical history. An integrated biopsychosocial assessment may help communicate the significant role that psychosocial factors play in the etiology and maintenance of somatic symptoms. Helpful prompts include, *"Do you consider yourself to be a worrier? Do you tend to get easily stressed out or down on yourself?"* (see Table 19.1). Positive responses to general prompts should be followed by more detailed assessment of situations and contexts in which these anxious or depressive symptoms tend to occur. Replicating the families' language (e.g., *worrier, cautious, hard on herself*) rather than using clinical terms (e.g., *anxious, depressed*) may facilitate an open discussion that will generate more detailed information.

Educating and Engaging Patients in CBT

Once physicians have determined physical symptoms are functional and that the child may benefit from CBT, it is important to carefully explain FSS to patients. Given that children and their families are seeking medical treatment for physical distress, families may become confused or defensive when a psychological treatment is suggested. Therefore, validating the physical

Table 19.1 Assessment prompts and questions

General prompts (can be altered to ask parent about child)
• *Do you put a lot of pressure on yourself or have really high standards?*
• *Would you describe yourself as overly cautious or careful?*
• *Would you describe yourself as shy or slow to warm up?*
• *Do you tend to be really hard on yourself?*
• *Would you describe yourself as overly negative or have you noticed that you get stuck thinking about things long after they happened?*
• *Do you tend to avoid situations that seem scary or overwhelming?*
Questions for assessing patterns of physical and emotional symptoms
Do you tend to have more pain/discomfort…
• School-related factors
– *On the weekdays vs. the weekends? On Sunday nights and/or Monday mornings? At the end of school breaks? In the mornings before school or at bedtime? During particular classes that you find difficult/stressful?*
• Achievement or performance situations
– *Around times when there are tests/projects/presentations (*e.g.*, the night before)? When preparing for or prior to athletic or musical performances?*
• Bullying/peer rejection
– *Is anyone giving you a difficult time at school? Do you tend to have [child's physical symptoms] when anticipating seeing those peers?*
• Social situations
– *When going out with friends, going to parties, or making plans? Waiting to be invited out?*
• Separation from caregivers
– *When leaving parents (or care givers)? When having to sleep away from home like sleepovers or camp? When parent is away on a trip? When having to sleep in own room?*
• Novel situations (e.g., field trips, changes in schedules/plans)
• Family disruption
– *Have there been any changes at home lately?* If parents are separated/divorced: *Any changes in custody arrangement? Has there been an increase in arguing at home? Changes in who takes care of you and when? Any recent deaths or losses?*

symptoms and providing education about the biopsychosocial model may improve families' acceptance and pursuit of nonmedical interventions such as CBT.

Validating Functional Somatic Symptoms

Perhaps the single most important thing the physician can do to facilitate effective treatment of FSS is to validate that the physical pain is real and distressing. For example, "*The good news is that we have not found anything positive on all the tests we have done. The bad news is that you are having pain and we have to try and figure out a way to help you feel better and get back to doing the things you like to do* (see Table 19.2)*.*" Expressing relief rather than confusion when indications of organic disease, such as potentially "dangerous" symptoms (e.g., blood in stool, disruptions in growth, abnormalities in blood chemistry or brain imaging), are absent can reinforce this message. In summary, the physician's reaction can help prevent the family from interpreting negative medical findings as, "*There is nothing physically wrong so it must be all in my head. I must be crazy*."

Psychoeducation: Explanation of the Biopsychosocial Model

As a next step, psychoeducation about the biopsychosocial model, particularly the connection between emotional and physical feelings, is a critical factor in engaging families who may be skeptical of receiving a "psychological" intervention for their child's "physical" symptoms. Families' attributions about the cause of FSS

Table 19.2 Psychoeducation and normalization

This might sound surprising, but medicine is far from a perfect science, so just because the tests we've done haven't revealed a specific cause, doesn't mean that the symptoms that you are experiencing aren't real. While the tests I conducted don't tell us what is causing your symptoms, I can tell you with certainty what I have thankfully been able to rule out [insert organic diseases] *by the results of* [insert specific tests]. *Based on our conversation about how you tend to be really hard on yourself* (use family's language), *I suspect that stress/worry about* [insert triggers/situations they endorsed] *might be contributing to your symptoms*
Did you know that a lot of recent studies have shown that stress, worry, or feeling really down can cause physical symptoms similar to the ones you are describing? We don't know a lot, but we know that there are pathways from our brain to all parts of our body that can carry nervous chemicals and hormones. We have also learned that some people are more sensitive in certain parts of their system. Think about it as having nerves in your [insert specific body part, e.g., stomach] *with the volume turned up, so the sensations you feel are amplified. Other people start out with an infection or some other type of disease, but for some reason they remain extra sensitive once the original disease is healed. For example, have you ever gotten a burn or know someone who has? We often find that, even once the skin is "healed," the person still experiences sensations differently in that spot when doing simple things like putting clothes over the skin*
Since we feel confident that you do not have a serious medical condition, I think we should shift away from figuring out "why" and start focusing on what to do about your symptoms so that you can get your life back to normal. Part of our approach will be to continue to keep track of your symptoms and what seems to trigger them. I suspect that some of the [psychosocial] situations you described to me may be contributing to your physical symptoms. Monitoring your symptoms will help us figure out whether you need to work with a specialist who helps kids learn how to deal with stress/worry/pain/etc (use family's language), *or whether I want to do additional tests or refer you to a doctor who specializes in those kinds of symptoms*

have been theorized to be a critical factor for successful outcome [48]. A study of youth with recurrent abdominal pain showed that parents' belief that their child's pain was primarily caused by psychological factors differentiated children who recovered from those who continued to experience pain years later [49]. The biopsychosocial model posits that psychological factors (e.g., early life events, family, coping, trait anxiety), interact with genetic and physiological characteristics (e.g., motility, inflammation, sensation), to determine an individual's susceptibility to developing FSS [50]. It assumes that genetic background and early life experiences influence an individuals' predisposition to FSS.

The development of FSS depends on one's response to physiological stimuli, which is affected by physical, environmental, and social exposures. For example, fear of pain is thought to be an important mechanism underlying the development and maintenance of chronic pain. Specifically, experiencing discomfort can lead to fear of pain, which subsequently increases attention toward pain and the likelihood that one will avoid situations associated with discomfort, thereby increasing disability [51, 52]. Explanation of the biopsychosocial model provides families with a general understanding of the etiology of FSS that supports the use of a psychological intervention.

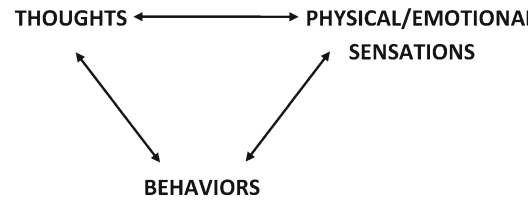

Fig. 19.1 CBT triangle

Psychoeducation: The CBT Model

Describing the CBT model is another important strategy because it addresses apprehension about how a psychological intervention will alleviate the child's distressing physical symptoms. The CBT model purports that complex and reciprocal interactions exist between physical and emotional feelings, thoughts, and behaviors (see Fig. 19.1). CBT focuses on directly changing thoughts and behavior in order to indirectly affect emotional and physical feelings. For example, thoughts such as, *"I'm going to get a stomachache while I'm at the party and everyone will notice,"* can increase feelings of anxiety, which can bring on

or exacerbate physical sensations like stomach pain. The stomach pains may then lead to avoidance, for example, deciding to skip the party. While such avoidance decreases distress in the short term, the relief experienced by avoidance helps to maintains anxiety and functional symptoms in the long run. Similarly, physical discomfort (e.g., headaches) may lead to negative thoughts like, "*I must have some kind of serious disease,*" which can increase nervousness or sad feelings, leading to withdrawal or isolation. When explaining these concepts to families, using examples of physical symptoms, cognitions, and avoidance that are typical of the child's experience can strengthen families' understanding of these connections, which is crucial for creating buy-in that CBT interventions will address the child's specific set of symptoms.

Brief Cognitive-Behavioral Strategies for Use in Pediatric Medical Settings

The following cognitive-behavioral strategies, using monitoring to recognize symptom patterns and relaxation, may be feasible for implementation within the pediatric office if there is a professional (e.g., physician, nurse, or social worker) who can devote a few 15–20 min follow-up meetings to the family. For children with mild symptoms or a recent onset of symptoms, this may be preferable to providing an external referral. Families may be more willing to accept behavioral intervention delivered by a medical professional with whom they have a pre-existing relationship.

Recognizing Symptom Patterns Through Monitoring

Identifying patterns that highlight common physical–emotional connections in the child's daily lives (e.g., stomach pain often occurs during anticipation of an anxiety-provoking situation) can help bolster families' acceptance of the biopsychosocial model and rationale for a psychological intervention. Asking families to keep daily written logs of physical discomfort and feelings (e.g., stress, worry, feeling down) helps to emphasize these interactions. First, patients should be instructed to assign a quantitative value (e.g., 0–8) for emotional and physical symptoms and be instructed to record these ratings as well as what activity or behavior they were doing or anticipating at the time (see Fig. 19.2). For children and adolescents with food-related concerns, a column listing food intake should be added to further explore this relationship.

During follow-up visits, the physician or medical professional should review these monitoring forms with the family to highlight the emotional–physical connections and to identify situations that tend to be associated with the onset of physical symptoms. Patterns characteristic of youth with FSS include discomfort associated with going to school, achievement or performance situations, social interactions, separation from caregivers, and novel situations (see Table 19.1 for specific situations). Additionally, youth with persistent discomfort may develop fear or avoidance in anticipation of experiencing the physical symptoms (e.g., being away from a private bathroom, long car trips, eating certain foods, exercise, loud stimuli), which can exacerbate somatic symptoms and maintain avoidance.

For some youth, gaining a better understanding of their symptoms through monitoring may decrease negative attributions (e.g., "*There is something medically wrong with me that the doctors are missing*"), which sometimes results in symptom relief. However, many children will require additional intervention, as described below. To determine whether improvement has occurred, any parent- or self-report measures administered during the initial evaluation should be repeated. Alternatively, comparing patients' monitoring forms to earlier time-points can inform decisions about whether more intensive CBT techniques are warranted.

Relaxation

Given that many types of relaxation strategies will be covered in other chapters of this book, we will not discuss this technique at length. Strategies typically used in CBT include diaphragmatic breathing and thematic imagery (Chap. 22), which can easily be implemented by a physician or other medical professional without extensive training. The literature supports the use of relaxation strategies for reducing the severity and

Fig. 19.2 Symptom monitoring form

frequency of headache and abdominal pain in children and adolescents [37–40], but relaxation may not be sufficient for reducing internalizing symptoms in isolation from other more specialized CBT strategies such as exposure.

Cognitive-Behavioral Strategies Requiring Training

Cognitive restructuring, exposure, and behavioral activation are more complex CBT techniques that physicians and medical professionals can implement with ongoing consultation with a specialist who has received specific training in these strategies.

Cognitive Restructuring

Children with FSS tend to negatively evaluate their ability to cope with pain and exaggerate the negative consequences of experiencing physical discomfort in public. These exaggerated or catastrophic thoughts are theorized to have a significant impact on the development and maintenance of internalizing symptoms. A study of clinically referred adolescents with chronic pain showed that catastrophizing was one of the most powerful predictors of anxious and depressive symptoms [53]. In addition to exacerbating internalizing symptoms, negative thoughts also contribute to avoidance of social and physical activities, and are therefore a main target of intervention for adolescents and older children.

Cognitive restructuring, or realistic thinking, teaches youth to identify negative predictions in order to challenge and replace them with more realistic cognitions. So instead of thinking, *"I can't go to the party because I'm going to have stomach problems and everyone will notice,"* youth can learn to replace these thoughts with more realistic ones, like "*If I start to feel sick I can excuse myself and go to the bathroom,"* or *"Everyone needs a bathroom break sometimes. No one will think that is weird."* Helping youth to think more realistically can create a greater sense of confidence in their ability to face feared situations

and encourage gradual reengagement in activities they have been avoiding.

Exposure

Many families dealing with persistent FSS and anxiety have fallen into the trap of avoiding situations that are associated with physical discomfort, likely because it is an effective strategy for decreasing symptoms in the moment. As mentioned above, however, avoidance tends to reinforce catastrophic thoughts about the child's inability to cope with the situation and ultimately leads to further avoidance because the immediate decrease in anxiety functions as negative reinforcement. Therefore, the first step in breaking this cycle is educating families that repeatedly approaching situations that provoke anxiety or physical discomfort actually decreases anxiety over time [54]. While approaching situations may not directly decrease physical symptoms, it may affect these symptoms indirectly through reducing the anxiety associated with situational triggers. It should be pointed out that experiencing pain while engaging in necessary (e.g., school) or preferred (e.g., out with friends) activities may improve the child's ability to tolerate pain and may decrease perceived pain by distracting youth from the pain. Overall, the message must be conveyed to families that although it may seem counterintuitive, approaching rather than avoiding anxiety- and pain-provoking situations will ultimately decrease these distressing symptoms.

As might be expected, this is not always a well-received recommendation and requires a great deal of validation that the clinician understands how difficult it will be to approach these situations. We often frame facing fears as an experiment (e.g., "*I understand that this is scary, but would you be willing to do it and see what happens?*"). Another critical message for preparing youth to engage in exposures is to convey that they will have input in which situations they will be targeting. Hierarchies of feared (but not dangerous) situations are collaboratively developed by ranking situations evoking mild anxiety, which should be addressed first, to those associated with severe anxiety, which must be approached gradually. For children with FSS, typical feared situations include being away from a (private) bathroom, eating certain foods associated with physical discomfort, being in public while experiencing physical discomfort, and exerting physical activity.

Exposure typically includes practicing situations with the family in the office to ensure that they understand the procedure and that the child experiences some success, in addition to assigning practice homework exercises for naturally occurring feared situations in the family's everyday life. Following in-office or homework exposures, it is useful to evaluate whether the "experiment" was better than the child expected, how much physical discomfort they experienced, and how well they were able to cope with it. Exposures allow youth to learn through experience that their anxiety will decrease over time, that anxiety is temporary and not dangerous, that negative predictions are unlikely to be realized, and that they have the ability to cope with uncomfortable situations [54].

Behavioral Activation

Behavioral exercises may also target avoidance that is driven by depressive symptoms such as fatigue or loss of pleasure rather than fear. Behavioral activation, an intervention that has received support for youth with depressive disorders [36], aims to replace avoidance, withdrawal, and inactivity with alternative coping, specifically graded "approach" behaviors. This includes establishing routines and encouraging youth to engage in activities previously experienced as pleasurable in order to increase the probability of receiving positive reinforcement from the environment [55], increasing the likelihood that the child will feel motivated to continue engaging in these activities. Examples relevant to youth with FSS and depressed mood include creating an exercise routine, developing a list of simple activities that the child can engage in when feeling down, or scheduling outings with peers or family on a regular basis (e.g., once per weekend). Additional applications of behavioral activation are described in the second case study presented below.

Making Referrals for Specialized CBT

When implementing cognitive-behavioral strategies within the physicians' office is not feasible, or if symptoms persist beyond a few follow-up appointments, more intensive application of CBT techniques delivered by a specialist might be indicated. An immediate referral is also warranted when moderate to severe anxiety or depressive symptoms are present and causing significant functional impairment, such as school refusal, avoiding important or desired social activities, impairment in school performance or friendships, difficulty falling or staying asleep, or significant food-related anxiety. When referral is necessary, all of the engagement strategies discussed above can be very useful, particularly reminding patients of the biopsychosocial model and patterns that were identified through monitoring.

Normalizing the Referral

Given the stigma surrounding mental health treatment [56], the manner in which the referral is presented may play a critical role in increasing the likelihood that families will pursue CBT. It is important to communicate that a referral for CBT does not indicate that the physical symptoms are not "real," but rather that changing behaviors and thoughts can have a "real" effect on the body's physical response. It may be helpful to cite scientific evidence, for example, that research has shown medication, dietary changes, and supplements to be largely ineffective [29–32]. Families should also be reminded that medication can have adverse physical side effects, which may worsen somatic distress. Finally, it is important to explain to families that CBT is different from how they may think of "psychotherapy." Specifically, CBT is action oriented, with patients practicing realistic thinking and approaching real world situations that they have been avoiding or find distressing. Parents and children are always involved in choosing the treatment goals and designing practice exercises.

How to Find a CBT Practitioner

Difficulty finding a CBT practitioner and knowing how to evaluate therapists can be a potential barrier to following through with a referral. Providing a list of appropriate questions for families to ask therapists to determine whether they specialize in CBT may be helpful (see Handout in the Appendix). A good resource for locating CBT therapists is the directory on the Association of Behavioral and Cognitive Therapies Website (http://www.abct.org). Once a referral has been made, it may be helpful to follow-up with the family in a few months. This may communicate to the family that their physician is invested in a successful outcome, may make them more accountable in following through with the referral, and may allow for feedback about CBT practitioners in the area. Establishing consulting relationships with qualified CBT therapists may expedite the referral process for future patients and facilitate ongoing communication regarding shared patients.

Case Studies

The following case studies illustrate the CBT strategies presented above. The first case, Sarah, provides an example of the brief CBT for FSS and anxiety using psychoeducation, monitoring, and education about approaching feared situations. In this case, it was feasible for these strategies to be implemented within the physician's office. The second case, Andrew, illustrates persistent FSS with comorbid anxiety and depression and associated sleep difficulties. Given the severity and persistence of symptoms, Andrew was referred to the pediatric social worker affiliated with his pediatrician. This case demonstrates specialized components of CBT delivered by a social worker with training in CBT, including realistic thinking, exposure, and behavioral activation.

Case Study 1: Sarah

Sarah, an 11-year-old Latina female, had a viral infection 3 months ago. Following her infection, she complained of daily nausea, gas, and abdominal pain. At the consultation meeting with her pediatrician, Sarah appeared anxious (e.g., clung

to her mother's hand, fidgeted) and became tearful when her mother left the examination room. It was also noted that Sarah's mother asked for repeated reassurance about her parenting skills, often related to situations in which Sarah became upset. Based on the physical symptoms reported, the family was referred to a pediatric gastroenterologist for further evaluation, who, after an endoscopy, ruled out medical pathology.

As Sarah's physical symptoms persisted, the family returned to their pediatrician in hopes of finding relief. During this visit, the pediatrician normalized the presence of Sarah's physical symptoms, validating their existence and the impact they had on her life. The role of stress on physical well-being (see Table 19.2) was introduced, followed by instructions of how to record Sarah's gastrointestinal discomfort, stress triggers, and food consumed for 2 weeks (see Fig. 19.2). At a follow-up meeting, it was noted that Sarah's gastrointestinal discomfort occurred around meals, with most intense discomfort associated with being away from home. Additional questioning (see Table 19.1) revealed Sarah experienced anxiety in anticipation of stomach symptoms in situations that she might not be able to locate a bathroom. Her worries were triggered by school, travel, and situations with unknown bathroom access or long distances to bathrooms (e.g., large parks, mall).

To educate the family, the CBT triangle (see Fig. 19.1) was introduced to demonstrate the interaction between thoughts, emotions, physical symptoms, and behaviors. For instance, the presence of Sarah's physical discomfort triggered thoughts that she had "no control over her body and may have an accident" and that there was something "medically wrong" with her. In turn, her thoughts then contributed to avoiding school in the morning, leaving class to visit the nurse, and distress on her way to events such as soccer practice where the bathroom was not easily accessible. The family was coached to help Sarah generate coping statements when experiencing gastrointestinal discomfort (e.g., *"I am medically ok even though I have physical discomfort," "I make decisions, not my pain"*) and was encouraged to go to school when she had gastrointestinal symptoms. This experiment would allow Sarah to test whether her predictions (e.g., having an accident) would actually happen.

In a second follow-up session a week later, Sarah and her parents reported success in using coping thoughts and, with the coaching of her parents, attending school one day despite feeling nauseous. She was nervous to go, but reported she felt better after 30 min, and acknowledged it wasn't as bad as she predicted. This exercise demonstrated that her stomach pain did not always lead to needing the bathroom, that it would eventually dissipate, and that she was able to tolerate discomfort in class. She had also discontinued visiting the school nurse as she no longer believed there was something "medically wrong" with her. Given this initial success, the family was asked to continue assignments with the goal of gradually attending school on time every day before their next follow-up meeting in a month.

When the family returned, Sarah had made tremendous progress in attending school on time, although she continued to arrive late about once per week. When asked what was happening on those days, Sarah's mother reported doubts about pushing her daughter and endorsed feelings of guilt when her daughter became upset. These concerns were validated, and the disruptive nature of allowing avoidance was reviewed. Rather than nurture anxiety, Sarah's mother was encouraged to tolerate Sarah's anxious behaviors, as this would help her daughter in the long run. She was instructed to focus on Sarah's brave behavior (providing praise and support) when Sarah attempted to manage physical discomfort independently.

At the last follow-up meeting another month later, Sarah had completely resumed all school activities and reported minimal gastrointestinal symptoms. Sarah and her mother noted that the validation and education they received was most helpful in meeting their goals. Furthermore, the physician's questions about Sarah's nervousness increased the family's comfort with discussing emotional factors and encouraged Sarah to push herself to tolerate physical and emotional discomfort.

Questions

1. During the initial consultation meeting, which of the following was a sign that Sarah might be experiencing anxiety related to separating from her mother?
   (a) Sarah became tearful when her mother left the examination room
   (b) Sarah displayed clingy behavior during the appointment
   (c) Sarah's mother asked for a reassurance about her parenting skills when Sarah becomes upset
   (d) All of the above
2. Which of the following CBT techniques were *NOT* used in this case example?
   (a) Symptom monitoring
   (b) Cognitive restructuring
   (c) Behavioral activation
   (d) Psychoeducation
3. Which of the following factors presents the most significant barrier to further treatment success and/or put Sarah at greatest risk for a relapse?
   (a) Sarah's chronic GI distress
   (b) Sarah's positive relationship with her physician
   (c) Sarah's negative, exaggerated cognitions
   (d) Sarah's mother's guilt associated with pushing her to face anxiety-provoking situations

Answers

1. (d): All of the above. Becoming upset or anxious upon separation from a parent or caregiver, as evidenced by Sarah's tearfulness when her mother left the examination, is a possible sign of separation anxiety. Clingy behavior, or physical attempts at closeness with a parent or caregiver, such as holding on to a parents' hand, arm, legs, or clothing is another sign of separation anxiety. Finally, Sarah's mother's need for reassurance about her parenting response when Sarah becomes upset implies that she is unsure of what to do in separation situations. Given additional information provided in the case, one might assume that Sarah's mother is behaving in an overprotective manner, sending the message that separation really is scary, which may be exacerbating Sarah's anxiety about separation. Other signs of separation anxiety that physicians can be vigilant of is avoidance or fear of being alone, sleeping alone, attending sleepovers or other overnight activities like camp, and concern for the safety of caregivers when separated (e.g., frequent asking about the caregiver, texting or calling repeatedly, asking for a precise time when the caregiver will return, etc).
2. (c): Behavioral activation is a technique for treating depressive symptoms, particularly anhedonia and withdrawal, which do not seem present in Sarah's case. Behavioral activation aims to replace avoidance, withdrawal, and inactivity by introducing graded "approach" behaviors that will increase the probability of receiving positive reinforcement from the environment. Each of the other treatment approaches, symptom monitoring (a), cognitive restructuring (b), and psychoeducation (c), were used to treat Sarah's separation anxiety symptoms in this case.
3. (d): While Sarah's history of chronic GI distress and her negative cognitions pose some risk to Sarah's continued progress, the greatest barrier is her mother's guilt about pushing Sarah to enter anxiety-provoking situations, which was still present toward the end of treatment. Sarah has made significant progress but would benefit from continued support from her mother and other family members as she faces new challenging tasks in her daily life. Given the family's positive relationship with the pediatrician, which is an important strength, it may be useful for the physician to check in via phone with this family from time to time and remind Sarah's mother that approaching anxiety-provoking situations is the most effective way of ensuring that Sarah will continue to make progress rather than slip back into her pattern of avoidance.

Case Study 2: Andrew

Andrew, a 16-year-old Caucasian male, began experiencing chronic daily headaches approximately one year prior to presenting to his pediatrician. He had been experiencing mild but increasingly

symptomatic headaches since the start of 9th grade, 2 years prior, which coincided with a change of school, more challenging homework and self-induced pressure to attend an ivy-league college. Over the last year his headaches became more frequent, resulting in difficulties falling asleep and daily fatigue. After extensive medical examinations conducted by his pediatrician and a pediatric neurologist, including brain imaging, his headaches were suspected to be functional, and he was referred to the pediatric social worker affiliated with his pediatrician.

Further evaluation by the social worker revealed Andrew, a self-proclaimed "high achiever," was spending a disproportionate amount of time on homework each night (5–6 hours) due to excessive checking and neatness. Given the time required to complete his homework, he did not engage in afterschool extracurricular or social activities with friends on school nights. In addition to his academic concerns, Andrew had rigid morning and evening routines, avoided being late, and spent a lot of time on his hair and clothes in order to make a "good impression." His worry about grades and making a good impression was accompanied by sleep disruptions, muscle tension, daily fatigue, and concentration difficulties. Additionally, for the last year, Andrew began experiencing depressed mood, a decline in confidence and interest in activities, weight loss of 12 lb, and thoughts of worthlessness and passive suicidal ideation.

Based on his presentation, Andrew was experiencing comorbid generalized anxiety and dysthymia. Although it is not possible to determine causality, the history suggested that Andrew's headaches were connected to his anxiety about school performance. Given the unremitting nature of his anxiety symptoms, his headaches worsened, contributing to fatigue and greater anxiety, creating a self-perpetuating cycle. Furthermore, as is common with unremitting anxiety, depressive symptoms began [57].

As a first step, treatment focused on helping Andrew understand the connection between his physical symptoms, worry, and mood. Given his somatic symptoms, Andrew was also asked to monitor his anxiety and physical pain daily to reveal possible interactions between pain and anxiety (using Fig. 19.2). After a few weeks, patterns emerged showing that his headaches were closely tied to anticipatory anxiety about the start of the school week (e.g., Sunday night), as well as tests and school presentations. It also became clear that his anxiety and headaches were leading contributors to his fatigue, as he was often unable to fall asleep those nights. This insight helped Andrew begin to conceptualize his headaches as a physiological response to anxiety and helped him understand the interaction between his emotional and physical symptoms.

Because Andrew was engaging in catastrophic thinking about school and physical discomfort, which he felt little control over, cognitive restructuring was implemented. He was guided to elicit his automatic thoughts (e.g., *What was going on in your mind when you began to feel anxious?*) and began examining evidence for his thoughts. For instance, Andrew frequently worried that his "*headache would make [him] fail the exam.*" To examine the evidence for the accuracy of this thought, Andrew was coached to ask himself the following: "*How many times have you actually not done well or failed on a test when you had a headache? Have you been able to succeed while having a headaches or feeling tired in the past? What is the worst that can happen? Would you be able to live with that outcome?*" As Andrew was an A student, many of these questions suggested that in all likelihood he would succeed, thus his fears were excessive and most likely unrealistic. Alternative interpretations of his maladaptive cognitions surrounding perfectionism and pain were generated and his ability to cope with pain was reinforced.

Andrew was successful at generating alternative thoughts, leading to a decrease in distress and frequency of headaches, but he continued to spend an excessive amount of time on homework. Exposures were implemented to test the accuracy of his alternative thoughts and reduce checking behaviors. A fear hierarchy was created:

Description of exposure task	Distress (0–8)
Only check homework one time	7
Forget to turn in homework	6.5
Make a purposeful mistake on homework	6
Arrive to class 5 min late	5

Andrew was gradually exposed to items on his fear hierarchy until items elicited minimal distress. Additionally, when he caught himself ruminating, Andrew was instructed to create a "to-do list" and focus on aspects that he could control rather than aspects out of his control. This was done for pain as well as anxiety. For instance, if he had a headache he could engage in relaxation, take a shower, or take a walk.

As treatment progressed, Andrew's mood symptoms improved, but he continued to feel tired and isolated. In order to target his fatigue, an exercise plan was put into place. It was explained that regular exercise, although it seems counterintuitive, would reduce fatigue in the long run by strengthening his stamina and releasing endorphins. Furthermore, regular exercise would make him feel tired at bedtime, thereby decreasing the time it would take to fall asleep. A review of his diet demonstrated that he ate mostly carbohydrates and food with little nutritional value; thus, a balanced meal plan was created. To reduce isolation and increase pleasure, regular social activities were gradually introduced and local family excursions were planned.

After 14 sessions, Andrew and his parents reported substantial improvement in stress, mood, and pain. He spent no more than 3 hours a night on homework and felt more relaxed. Although he continued to have occasional headaches, he was better able to cope with them and was engaging in more activities.

Questions

1. Why is Andrew's habit of spending 5–6 h on homework a symptom of anxiety?
   (a) Andrew's tendency to spend excessive time on homework is a sign of his extremely high standards and perfectionism
   (b) Spending too much time on homework is not a sign of anxiety
   (c) Spending so much time on homework is Andrew's way of avoiding participating in extracurricular activities
   (d) Spending excessive time on homework is Andrew's way of avoiding his depressive cognitions
2. What factors led to Andrew's physician deciding to make a referral to a social worker rather than applying CBT techniques herself?
   (a) Andrew's somatic and internalizing symptoms are long-standing (present for several years)
   (b) Andrew was experiencing suicidal ideation
   (c) Andrew was experiencing a number of both anxious and depressive symptoms
   (d) All of the above
3. What was the importance of encouraging Andrew to exercise and eat more nutritious foods?
   (a) To help him lose weight
   (b) To counteract his fatigue and sleep problems
   (c) To counteract his headache pain
   (d) To help him feel more healthy

Answers

1. (a): Spending an excessive amount of time on homework is often a sign of anxiety. We know that Andrew has high expectations for himself and feels significant pressure to earn excellent grades so that he can attend an Ivy-league college. These perfectionistic tendencies are also supported by his need to repeatedly check his work and ensure neatness, which likely also adds to the length of time he is spending on homework. In other cases, spending excessive time on homework could be a way of avoiding participation in extracurricular activities, but there is no evidence in this particular case that Andrew avoids social situations beyond his concern about making a good first impression, which is more likely related to his high standards and perfectionism.
2. (d): All of the above. The most striking reason for referral to a social worker in this case is Andrew's suicidal ideation, which must be carefully and continually monitored by a professional experienced in suicide risk assessment. The fact that Andrew experiences many long-standing symptoms of anxiety, depression, and headache, make this a more complex case that would likely benefit from a professional who can devote sufficient time and resources to develop a treatment plan that will include many of the techniques described in this chapter to adequately address this degree of comorbidity.
3. (b): While improvements in diet and exercise might have additional benefits, the purpose for

prescribing these changes is to combat Andrew's fatigue and sleep problems. If Andrew can establish a regular exercise routine, one would expect that he might fall asleep more quickly at night because he will be more tired. In addition, regular exercise will help reduce fatigue by increasing his stamina. Changing Andrew's diet by decreasing carbohydrates and increasing nutritional value by introducing more protein should also help to increase his energy level throughout the day.

Conclusions

Functional somatic symptoms are common in youth and associated with persistent disability and repeated medical evaluation. Internalizing disorders often co-occur, yet they are rarely identified and mental health treatment is the exception. Recent research has shown promise for the use of CBT, a well-established intervention for anxiety and depression, for youth with FSS. Physicians play a critical role in facilitating appropriate treatment for these youngsters by identifying internalizing symptoms, validating the existence of pain without an explanatory organic disease, explaining the biopsychosocial model, and promoting acceptance and pursuit of psychological intervention. While additional research is needed to support the efficacy of CBT for youth with FSS and internalizing symptoms, psychological treatment for this population appears to be a viable option. The implementation of CBT strategies by professionals within the pediatric setting is preferable to providing external referrals due to the stigma associated with seeking mental health treatment, particularly for families that may continue to view FSS as a "medical" problem. Additional research is needed to develop and evaluate service delivery models that will support the implementation of brief evidence-based psychological intervention, such as CBT, within the medical setting. Identifying cost-effective strategies for training medical personnel in cognitive-behavioral strategies and developing service delivery models that alleviate the current logistical barriers that preclude mental health professionals from functioning within pediatric offices stand to improve clinical care of youth with FSS and internalizing disorders across pediatric settings.

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The Use of Biofeedback and Neurofeedback in Pediatric Care

20

Donald P. Moss

Abstract

Biofeedback and hypnosis are complementary techniques, sharing an evidence-based approach and accessing powerful neurophysiological mechanisms. Both approaches are suitable for use with children, when implemented in developmentally appropriate ways, and each facilitates the development of self-regulation. Biofeedback can serve as an adjunct to hypnotherapy, and hypnosis can augment biofeedback treatment. Biofeedback and hypnosis can serve as alternative pathways for patient care or can be integrated into a comprehensive mind–body intervention plan. Common uses for biofeedback in pediatric care include the following: mind–body and affective education, biofeedback-assisted relaxation, neurofeedback for attention deficit hyperactivity disorder, biofeedback for medical problems, and neuromuscular education. Mind–body education teaches the child to recognize the effects of the mind—thoughts and feelings on the body. Affective education teaches the child to recognize emotions and cope with emotional responses to life situations. Neuromuscular education retrains muscles to recover movement after injury or illness affecting motor nerve control or central nervous system control of the musculature. In addition, biofeedback is a useful tool for optimal performance applications.

Keywords

Pediatrics • Biofeedback • Hypnosis • Self-regulation • Integrative care

What Is Biofeedback?

Defining Biofeedback

Biofeedback is a training approach for modifying human behavior, neurophysiology, and states of consciousness. Biofeedback uses electronic instruments to measure physiological processes,

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by giving instantaneous feedback to an individual about his or her physiology. This instantaneous feedback is called "real-time" feedback. The feedback enhances the individual's *awareness* of his or her own bodily processes and facilitates the individual's ability to gain *voluntary control* over and *modify* this physiology. The simplest examples of biofeedback involve monitoring the muscle activity in a single muscle or a group of muscles and providing visual or auditory feedback to the person, who is then able to increase or decrease activation of that muscle. John Basmajian [1] pioneered this process of muscle biofeedback in the 1950s and 1960s and showed that human beings, with feedback, are capable of controlling even a single motor unit.

Scientific Origins of Biofeedback

Biofeedback was based on laboratory research studies applying operant conditioning to animals and humans. Early biofeedback research showed the possibility of voluntary control, through feedback, of visceral physiology [2], blood pressure [3], and brain activity [4]. By 1969, basic research on human self-regulation of physiology advanced to a point that a scientific meeting was held in Santa Monica, California, to name and promote this new approach to human behavioral change. The name *biofeedback* won out over several other ideas, such as "autoregulation." From the beginning, this field has brought together the lab scientists and those dedicated to higher states of human consciousness. As Joseph Kamiya has described it, the white lab coats of the scientists came together with the white robes of the gurus (personal communication).

Neurofeedback

Neurofeedback is a branch of biofeedback and specifically applies the feedback paradigm to the training of brain activity, based on the early work of Kamiya [4], Fehmi [5], and Sterman [6]. Neurofeedback is utilized to remediate specific abnormalities in brain activation patterns, correlated with clinical disorders, including attention deficit hyperactivity disorder (ADHD) and convulsive disorders. Neurofeedback is also used to introduce altered states of consciousness facilitating the resolution of past traumatic experiences [7] or enabling greater creativity and optimal performance [8].

Biofeedback Modalities and Instruments

Biofeedback is a process that enables an individual to learn how to change physiological activity for the purposes of improving health and performance. Precise instruments measure physiological activity such as brain waves, heart function, breathing, muscle activity, and skin temperature. These instruments rapidly and accurately "feedback" information to the user. The presentation of this physiological information—often in conjunction with changes in thinking, emotions, and behavior—supports desired physiological changes. Over time, these changes can endure without continued use of an instrument.

A complete overview of the modalities used in biofeedback and neurofeedback practices is available in Shaffer and Moss [9] or in Schwartz and Andrasik [10]. Table 20.1 shows the major biofeedback modalities, including the instrument, the physiological process measured, the sensor, and the commonly used unit of measurement.

Progress in biofeedback since 1969, when the term biofeedback was coined, has followed the development of physiological measurement devices. To a large degree, if the technology can be developed to measure a physiological process, the human being can develop some voluntary control over that process. The best example is heart rate variability (HRV). Heart rate changes and patterns in heart rate change have been significant for medical research for 30 years. But only in the past 15 years have computer-based HRV instrumentation systems made HRV training practical for the average clinician. In this brief span, multiple research studies have measured a positive therapeutic benefit for HRV training, for asthma [11], fibromyalgia [12], depression [13], functional abdominal pain [14], and posttraumatic stress disorders [15]. The next wave in biofeedback

Table 20.1 Major biofeedback modalities, reprinted with permission

Modality	Acronym	Activity measured	Sensor	Measurement unit
Electromyograph	EMG	Muscle action potentials	Precious metal or post	Microvolts (mV)
Feedback thermometer	TEMP	Peripheral blood flow	Thermistor	Degrees F or C
Infrared thermometer	TEMP	Peripheral blood flow	Infrared detector	Degrees F or C
Photoplethysmograph	PPG	Peripheral blood flow, heart rate, heart rate variability	PPG sensor	Arbitrary units
Electrocardiogram	EKG	Heart electrical activity, heart rate, heart rate variability	Precious metal	Beats per minute
Electrodermograph	EDR, GSR, SCL	Eccrine sweat gland activity, electrical conductance/ resistance in skin	Zinc or precious metal	Microsiemens (mS)
Electroencephalograph	EEG	Cortical postsynaptic potentials	Precious metal	Microvolts (mV)
Pneumograph	RESP	Abdominal/chest expansion	Strain gauge	Arbitrary units
Capnometer	CAP	End-tidal CO_2	Infrared detector	Torr

Reprinted from Shaffer and Moss [9]. With permission from Taylor & Francis Books

development now lies in utilizing neuroimaging techniques, such as fMRI imaging, to facilitate voluntary control over more specific subcortical brain processes [16].

Biofeedback and Hypnosis

Biofeedback and hypnosis are two approaches to facilitate enhanced self-regulation, alleviation of clinical distress, general wellness, and optimal performance. Both approaches draw on the healing resources within the human being as a neurobiological organism. Both approaches are evidence-based, drawing on thousands of outcome studies documenting clinical efficacy and effectiveness, for a broad range of medical and emotional disorders. Both biofeedback and hypnosis induce transformations subjectively, in neural activation patterns and in general physiology. The evidence base for these broad statements will be presented in the remainder of this chapter.

Accessing Neurophysiology

Hypnosis and biofeedback are both neurophysiological in effect. When Linda Thomson, a leader in pediatric hypnosis, encourages a child with Tourette's syndrome to go inside himself, "find his tic switch," grasp the tic switch, and "turn it down, way down," the child is drawing on creative imagination but is also accessing neural pathways to reduce tic activity [17]. When Lynda Thompson, a leader in pediatric neurofeedback, utilizes neurofeedback to reduce the hyperactivity and inattention in a child with ADHD, she is also drawing on neural pathways but in a direct measurable fashion. The child observes visual displays communicating current cortical activation and learns to directly increase brain activation in the low beta range and reduce theta range activity, at sites monitored during the session [18].

Biofeedback as an Adjunct to Hypnosis

Biofeedback or "physiological monitoring" can serve as an adjunct to hypnotherapy (Chap. 21). The practitioner begins by monitoring the child's heart rate, skin conductance, respiration, or even brain wave activity during hypnotherapy sessions. Both the therapist and the child will begin to notice signs in the biofeedback line graphs and screen displays when topics in therapy induce stress and anxiety. Similarly, when a child responds effectively to hypnotically induced relaxation, the therapist can point to the computer display to show the child how effectively the hypnosis is calming his or her physiology. "Calming the physiology" can include reducing muscle activation, slowing respiration, slowing heart rate, reducing blood pressure, and in general reducing the effects of stress and the sympathetic

nervous system's response to stress on the body. Children are often more motivated for hypnosis when they see such physiological "proof" of the impact of hypnosis.

Linda Thomson [19] has advocated the strategy of teaching the child that hypnosis can help him or her to become the "boss of my body." Biofeedback devices concretely show the child that he is or she really is the *boss*, because actions taken by the child produce visible changes in the physiological signals.

Biofeedback sessions can improve hypnotizability and even serve as a specific induction technique [20]. This author has observed that many children who initially have seemed ill at ease or respond poorly with hypnosis often find the use of suitable biofeedback devices attractive and approachable. After three to four sessions of biofeedback, the same children are often more amenable to using hypnosis. I frequently use the child-friendly emWave™ HRV system from HeartMath, with its animations and games. After brief use of the emWave, many children seem more susceptible to trance induction.

Hypnosis as an Adjunct to Biofeedback

Similarly, hypnosis and hypnotically induced relaxation are useful adjuncts to assist children in their biofeedback training. For example, when a child is having difficulty mastering thermal biofeedback—warming of the hands and fingers, associated with autonomic nervous relaxation—a hypnotic induction and use of imagery of a warm beach or a fireplace can provide a breakthrough and empower the child. Similarly, self-hypnosis skills can be integrated into biofeedback home practice, to augment the child's emerging relaxation skills.

Biofeedback and Hypnosis as Options in Treatment Planning

Ian Wickramasekera [21, 22] promoted a model for selecting either hypnosis or biofeedback for a given patient, based on the individual's hypnotizability. For individuals who are high in hypnotizability, hypnosis represents a fast track for brief intuitive interventions producing dramatic physiological and emotional transformation. In turn, when an individual is low in hypnotizability, biofeedback is often effective, providing a concrete step-by-step change process, guided by visible physiological signals, and assuring the individual of real changes. Pragmatically, biofeedback offers a viable alternative for patients who are resistant to hypnosis for religious reasons or due to fears based on cultural stereotypes about hypnosis. Biofeedback can serve to guide the patient into deeply relaxed states, giving access to many of the same dynamic transformations that are commonly sought in hypnosis-based psychotherapy.

The Integration of Biofeedback and Hypnosis

Many practitioners have advocated integrating biofeedback and hypnosis in pediatric treatment [20, 23–27]. Timothy Culbert and colleagues have portrayed biofeedback and hypnosis as complementary approaches, easily blended, and each facilitating the development of self-regulation in children. In combination, the two strategies assist the child to cultivate low arousal states, enhance the child's focus of attention, deepen awareness of internal events, facilitate the use of imagery and fantasy, develop an internal locus of control, foster both empowerment and self-awareness, reinforce awareness of the mind–body linkage, enhance access to unconscious "material," and heighten suggestibility [20].

Biofeedback and Children

Children are ready subjects for biofeedback, although biofeedback techniques have to be adapted to the developmental age of the child participating in training. Research supported early in the history of biofeedback that children often master biofeedback tasks more effectively than adults [28]. Several leading figures promoted

the application of biofeedback with children, alone and in combination with hypnosis. Karen Olness [29], as well as Timothy Culbert, Jud Reaney, and Daniel Kohen [20], has promoted the use of biofeedback and hypnosis as cyber-physiologic strategies that facilitate emotional and physiological self-regulation in the child. The term cyber-physiologic derives from the Greek *Kyber*, meaning to take the helm or steer. Thus, a cyber-physiologic strategy teaches the child to direct or steer his or her own physiology, through establishment of voluntary control over bodily processes.

Children are often enthused and delighted to discover that they can control aspects of their body, which previously were troublesome. They often succeed in modifying their physiology easily and quickly compared to adults [28]. Today's children and adolescents are also technology-friendly, so doing something on the computer is often welcomed and perhaps exciting for children.

On the other hand, children possess shorter attention spans than adults, become bored more quickly, and may need fairly stimulating forms of feedback to sustain their engagement in the treatment process. Fortunately, the designers of biofeedback instrumentation have been creative in embracing electronic innovations and electronic games to provide forms of feedback that sustain children's attention. In the early years of biofeedback, the displays sometimes consisted of toy trains or "racing cars, linked in a relay with a biofeedback instrument, so that the train or car continued to move as long as the child relaxed a muscle below a threshold, warmed his or her hands above a threshold, or met some other designated physiological goal.

Today, biofeedback instrumentation systems are typically interfaced with a computer, and much more elaborate and entertaining forms of feedback are possible. For example, many biofeedback systems are programmed to control a DVD player so that the child can keep a movie playing or brighten the picture by reaching a training threshold or moving toward a training goal. Similarly, a variety of computer animations are available, so that a child can make a skeleton walk, drive a roller coaster, or complete a computer puzzle by correctly managing his or her physiology.

Efficacy of Pediatric Biofeedback

Four of the best documented applications of biofeedback in children are pediatric headache [30–32], attention-deficit disorder [33], gastrointestinal and eliminative disorders [34], and asthma [11, 35]. The body of research on biofeedback applications with children is relatively smaller and weaker than the research evidence on biofeedback for adults [34]. Benore and Banez [36] conducted a PubMed search and found only 222 articles published on biofeedback and children from 2008 to 2012, and of these only 36 were clinical trials, primarily focused on biofeedback applications with cerebral palsy, ADHD, and elimination disorders. Yucha and Montgomery [37], who have published systematic efficacy ratings on biofeedback and neurofeedback interventions, reported that treatment with these interventions can be rated as "probably efficacious" or "efficacious" for children with only a handful of clinical disorders—anxiety, ADHD, chronic pain, diabetes, epilepsy, fecal disorders, headache, insomnia, and traumatic brain injury. No pediatric application met the highest rating of "efficacious and specific," largely because of the lack of independent randomized controlled trials.

Nevertheless, a number of credible research reports show emerging promising applications of biofeedback for additional disorders, including functional abdominal pain [14, 38], childhood anxiety [39, 40], and autistic spectrum disorders [41–43].

Common Pediatric Uses for Biofeedback

In the following sections, we will discuss several of the most widely used pediatric applications of biofeedback.

Mind–Body Education

Biofeedback is an effective tool for teaching children to recognize the connection between body and mind and between emotions and the body. Many children suffer frightening physical symptoms in response to family and school stress, ranging from abdominal pain to headache to anxiety with racing heart. Children often lack the self-awareness to recognize the connection among thoughts, fears, and physical symptoms and frequently are resistant to talking about the related emotions. The practitioner can utilize biofeedback instruments to monitor heart rate, EEG, electrodermal response, muscle tension, or other physiological processes and display the results on a computer screen. Children are typically fascinated by this opportunity to learn about what is happening in their bodies. We can then invite the child to picture a stressful situation, or a problem he or she is currently facing, and show the child the impact of stress on the body. Then, we can guide the child to relax and view the impact of relaxation on the body, often reversing the previous effects of stress.

As long as the practitioner chooses only one or two physiological modalities to monitor and keeps the display simple, children will rapidly learn to recognize what the computer display is telling them about their bodies. Then, the therapist can engage the child in free ranging conversations about school, home, relationships, and problems or engage in drawing or play. When the computer display shows some kind of physiological reaction to the discussion, this presents an additional learning opportunity. We can simply invite the child to think about what we are discussing, or what he or she has been playing out with the puppets, and ask, "Why do you think that talking about your visits to your father make your muscles tense (or your heart go so much faster)?" We can also make interpretive comments, by using the biofeedback display to help the child see and accept the connection, for example, "Every time your voice gets real angry, your heart rate goes very fast. Did you realize your feelings were affecting your body so much?"

Biofeedback-Assisted Relaxation Therapy

The most common use of biofeedback with children is relaxation training. Some children will implement relaxation exercise instructions rapidly without any biofeedback, slowing their breathing, reducing tension in the musculature, and warming their hands. But for many, relaxation is a challenge. In some cases, family and life stress are so chronic and habitual that the child no longer feels or recognizes the accompanying physiological tension.

Biofeedback instruments offer a useful means to assist the child to relax. For example, we can show the child the course of his or her breathing, the pace of breathing, the sighs, the breath holds, or the irregularity of breathing on a line graph on the computer. Then, we can utilize a breath pacer and ask the child to "breathe in when the ball goes up and out when the ball goes down." This simple exercise will relax most children so much that they are surprised and will speak of "feeling sleepy."

Biofeedback instruments today, especially the systems that utilize software for a computer interface, are extremely flexible in their display capabilities, which is especially useful for children. We can "increase the gain," adjusting the unit of measure on the screen, so that hand temperature changes of 1/100th of a degree are visible on a bar graph, and these very small changes can trigger the onset of soothing music or cause a light bulb animation to glow. In this way, we can provide success experiences to children making very small physiological changes and in this way gradually enhance their self-efficacy, that is, their sense of an internal locus of control.

Current biofeedback systems also provide flexibility in the choice of display. The popular emWave™ system allows a child to colorize a garden and introduce flowers, a pony, and a butterfly or make a hot air balloon soar over a landscape, by breathing evenly and smoothly, thus causing heart rate variations to also smooth. Or for the "little scientist," who thrives on seeing the actual data, we can provide digital displays of the current muscle tension and a line graph with raw data.

"Feedback learning" is an important part of biofeedback-assisted relaxation therapy. When children can see their physiology in a computer display, they become more aware of their own process in raising or lowering hand temperature, tensing or relaxing muscles, or smoothing heart rate. If we allow the child to "play with the feedback" and provide clear simple feedback displays, children will almost always gain spontaneous control over the physiology in question. Many therapists emphasize teaching children imagery and specific relaxation skills and use the biofeedback display primarily to show the child when the relaxation skill is having the desired effect. This is a useful strategy, but therapists should not overlook the value of simple "feedback learning." Providing a clear feedback display increases awareness and the awareness facilitates control.

Neurofeedback for Attention Deficit Hyperactivity Disorder

Neurofeedback for ADHD is one of the most rapidly growing applications of biofeedback today. ADHD is a serious problem disrupting children's school performance as well as their social adjustment and extracurricular activities. Inattention and impulsiveness impede the child's success in the classroom, on the soccer field, on the playground, and throughout everyday life. ADHD is also in many cases a lifelong problem that persists into adult years, disrupting success in relationships and work.

Many parents are motivated today to seek alternatives to pharmacotherapy. Some children experience adverse affects from stimulant medication; others show only a moderate improvement in ADHD symptoms on medication. Long-term compliance with medication for ADHD is poor, and neurofeedback provides a clinically effective alternative.

Joel Lubar initially developed the neurofeedback training protocol for ADHD. He began with Barry Sterman's laboratory research on cats, commencing in the late 1960s, showing that training of the sensorimotor rhythm (SMR) over the brain's sensorimotor cortex led to a reduction in motor activity in cats. Sterman utilized these findings as the basis of developing a neurofeedback treatment for humans with seizure disorders [6, 44], a form of brain dysregulation.

Joel Lubar [45] extended this research to hyperactive children, recognizing that their disorder also involved brain dysregulation. Initially he trained hyperactive children to increase their brain's SMR activity, that is, electrical activity in the frequency range of 12–15 Hertz (Hz), and found a reduction in hyperactive behavior. Later he [46] discovered that training the child with ADHD to produce more brain activity in the lower beta range (14–18 H) and also to suppress slower activity in the theta range (4–8 Hz) enhanced attention as well. In additional studies, Lubar was able to show that successful training following this protocol produced improvements measurable on the Test of Variable Attention (TOVA) and significant improvements in overall IQ scores on the Wechsler Intelligence Scale for Children (WISC-R) [47]. Lubar [48] has conducted longer term follow-up studies, showing that the gains from neurofeedback persist for more than a decade in most cases, requiring only an occasional follow-up training session to restore optimal brain function. Monastra et al. [33] reviewed the research accumulating on the neurofeedback treatment of ADHD and rated this intervention as "efficacious" for alleviating the symptoms of ADHD. In 2012, the American Academy of Pediatrics published a ranking of evidence-based child and adolescent psychosocial interventions and included biofeedback for ADHD among the "Level 1 Best Supported" interventions [49].

Today the neurofeedback treatment for ADHD has evolved significantly [50]. A large number of children respond positively to the two simple Lubar protocols, initially training an increase in SMR activity to reduce any conspicuous hyperactivity and then training an increase in low beta (with a simultaneous reduction in theta) to enhance attention. However, additional subtypes of ADHD have been identified, based on diverging patterns of brain dysregulation. As a result, leading researchers advocate that neurofeedback

for ADHD should begin with a quantitative EEG, a brain map showing specific abnormalities in this child's cortical activation patterns [50]. Neurofeedback treatment then should be individualized to normalize brain activity for this specific child and his/her brain patterns [48]. Thompson and Thompson [18] have published guidelines for clinical use of neurofeedback for children.

Biofeedback for Medical Problems

Biofeedback has been applied to a wide variety of medical problems in adults, including asthma, diabetes, fibromyalgia, irritable bowel syndrome, hypertension, Raynaud's disease, and temporomandibular disorders [36]. As mentioned earlier, there is a lack of adequate well-controlled research on pediatric uses of biofeedback for medical problems. In this section, we will briefly examine two areas, asthma and headache, in which research is available.

Asthma

Asthma is a significant health problem for children, with an incidence increasing rapidly in recent years. According to the Centers for Disease Control, in 2010 an estimated 13.1 million children in the USA had been diagnosed with asthma, of which seven million had active asthma symptoms [51]. The disorder disrupts children's everyday lives, leads to missed school days, and causes a small but unacceptable numbers of deaths (0.3 deaths per 10,000 children with asthma) [51].

A variety of biofeedback and self-regulation approaches have been used with asthma [11, 52]. Early research used biofeedback for respiratory sounds and wheezing, with small positive results. Other studies showed modest improvements in asthma from relaxing the facial muscles, using frontalis (musculature of the forehead) surface electromyographic (SEMG) biofeedback. In addition, a number of studies used relaxation training without biofeedback, producing some significant but inconsistent outcomes [53].

The most promising biofeedback approach to date has been HRV biofeedback training [11]. Children with asthma tend to have low 24-h HRV during asymptomatic periods, apparently reflecting their body's diminished adaptive capacity [54]. In HRV training, patients learn to increase HRV by breathing at a specific rate, described as their "resonance frequency." This is the breathing rate which produces the highest variability in heart rate. For most people this is a breathing rate of about 6 times/min. Several small studies and one large controlled trial have found clinically significant improvements in asthma using this method [11]. Results in the Lehrer et al. [11] study showed an improved pulmonary function, a reduced need for medication, and an improvement in one full level of asthma severity (using an American Thoracic Society rating). Biofeedback, especially HRV training, appears to offer hope for treatment of children with asthma.

Pediatric Headache

A recent worldwide review of published studies on the prevalence of headache in children (through age 19) showed that headache is a global problem for children, with approximately 58 % of children suffering headache [55]. The prevalence of migraine is approximately 7.7 %. Andrasik and Schwartz [30] reported earlier figures, showing a rising prevalence of headache with age, from 20 % at age 5 to 57–82 % of children ages 7–15. Children with headache frequently miss school, withdraw from extracurricular activities, and at the extreme show significant disability, persisting into adulthood [56].

Biofeedback is well documented as an intervention for adult headaches, including both tension headache and migraine. The National Institutes of Health Technology Assessment Panel concluded that EMG biofeedback was superior to psychological placebo and comparable to relaxation therapies in treating tension headache [57]. The National Headache Foundation's *Standards of Care for Headache Diagnosis and Treatment* found that "biofeedback has been shown to be an excellent treatment in the long term management of migraine and tension-type headache disorders" [58].

Biofeedback has also been widely applied to children's headache problems, but the number of controlled research studies is relatively small [36]. Trautmann et al. [31] conducted a meta-analysis of biofeedback, relaxation training, and cognitive behavioral therapy for headache in children and found consistent evidence in 23 studies and case reports of improvement in headache in the children and adolescents treated. Their study provided support for psychological treatment of headache in children.

Muscle biofeedback (surface electromyography or SEMG) is commonly applied for tension-type headache, migraine, and mixed headache. In SEMG biofeedback, electrodes are placed on the forehead or cervical regions, and children are trained to reduce muscle tension, guided by a biofeedback display. Frequently, the child is also taught to use Edmund Jacobson's progressive muscle relaxation to assist in gaining muscle control. In progressive muscle relaxation, the child is guided to tense specific muscle groups, one at a time, feeling the sensations accompanying the tensing, and then to relax the same muscle group, feeling the contrasting sensations accompanying relaxation [59]. The combination of SEMG biofeedback and progressive muscle relaxation produces an enhanced awareness of muscle tension and significant gain in the ability to relax the musculature. In addition, children frequently come to recognize linkages between the onset of muscle tension and specific stressful life events, contributing to better coping.

Thermal biofeedback is commonly utilized for children with migraine and often for tension-type or mixed headaches as well. In thermal biofeedback, a sensor is placed on a finger, and the child learns to warm the finger, producing a dilation of the arterioles supplying blood to tissue. Children learn to warm their hands by direct biofeedback training, producing an animation and the onset of music each time the slightest increase in hand temperature commences. In addition, practitioners can guide children to use imagery of beaches, fireplaces, or their own toasty warm bed as aids to hand warming. In addition, autogenic training is a useful adjunctive technique to assist children in both hand warming and a deep autonomically based relaxation state [60]. That is, the relaxation is accompanied by significant changes in the autonomic nervous system, with a diminution of sympathetic nervous system activation and an enhanced recruitment of the parasympathetic nervous system.

Andrasik and Schwartz [30] provide a comprehensive and thorough review of the use of biofeedback for headache in adults and useful information for anyone treating headache in adults or children. They have advocated some modifications of the usual biofeedback procedures for use with children with headache. First, they remind the practitioner to attend to the child's developmental level, utilize appropriate language, and take the child's attention span into account. Second, they suggest including parents/family members in the treatment process, and discuss a "prudent, limited office treatment" approach, in which family members guide the learning of specific skills at home.

Neuromuscular Reeducation and Rehabilitation

Neuromuscular reeducation is the use of biofeedback for the restoration of motor control and functional movement, for children and adults affected by brain injury, stroke, neurological disease, and congenital conditions affecting neuromuscular function [61–63]. Neuromuscular reeducation involves a skilled analysis of the agonist and antagonist muscles needed to conduct a specific movement, which requires a thorough understanding of the musculature and kinesiology on the part of the practitioner. Then, the practitioner utilizes small surface electrodes with surface electromyographic biofeedback (SEMG) in order to record signals from specific muscles and trains constellations of muscles, inhibiting some and enhancing muscle recruitment in other muscles, to assist the child to carry specific actions. To gain a reward the child must respond in the targeted pattern.

When the child succeeds in making even small steps in the desired direction, some form of feedback is given as a reward. In many cases

today, a video plays or brightens and becomes more visible, each time that the child moves toward the training goals. This process of guiding the child in a stepwise fashion toward the combination of muscle actions needed for functional movement is called *shaping*. Shaping is one of many "operant conditioning" principles applied in neuromuscular reeducation [64, 65].

Bolek [66] conducted a retrospective review of the treatment of 16 children with various forms of neuromuscular deficits, at the Cleveland Clinic Children's Hospital for Rehabilitation. Each of the children had failed to respond to conventional interventions, including physical therapy and occupational therapy. The group included children with deficits in standing balance, sitting, head control, and upper extremity use. In each case, a training strategy was developed using SEMG to train a constellation of muscles, contracting some and relaxing others. Progress was mapped showing the percentage of time in each session that the child met the training criteria. The children were rewarded with videos each time they met the current training criteria. Verbal cues were also used along with the video reward, to assist *transfer* of the new skills outside the clinic setting. Fourteen of the sixteen children achieved a significant restoration of function.

Biofeedback-assisted neuromuscular reeducation presents a time-tested intervention to rehabilitate and restore neuromuscular function in children affected by a wide range of disorders, including cerebral palsy, stroke, accidental injury, and congenital conditions affecting motor function. Biofeedback-assisted neuromuscular reeducation can be delivered alone or in combination with physical therapy and occupational therapy. A case vignette will be provided in the Case Studies segment of this chapter, to illustrate the power of neuromuscular biofeedback to assist children who cannot be assisted by other means.

Pediatric Optimal Performance

Since the opening days of the biofeedback movement in the late 1960s and early 1970s, the biofeedback paradigm has excited visions of expanding human potential [67]. Biofeedback and neurofeedback are now widely used by practitioners in the field of optimal performance, to enhance human functioning in athletics and the arts [68–71]. There is a lack of controlled research on the application of optimal performance strategies with children and adolescents. However, practitioners have published a number of promising clinical reports and pilot studies.

Lagos and colleagues published a single case study showing the benefits of HRV for an adolescent competitive golfer. This 14-year-old maintained an average score of 70 during 18-hole golf practice, yet in competitions his average score worsened to 91. He suffered anxiety, with rapid heart rate, shortness of breath, and fear of losing control in competitions. After ten sessions of HRV training, his anxiety symptoms were diminished, his confidence increased, and his average score in competitions was reduced by 15 strokes [72].

Trechak [73] reported on her use of biofeedback and neurofeedback in optimal performance coaching with high school age musicians. She reported on her work with over 100 high school musicians, including both instrumentalists and vocalists; she also applied her approach with children affected by autistic spectrum disorders and assisted them to make both educational and artistic gains. Her work focused on reducing tensions, enhancing focus, and maintaining an optimal level of arousal.

Tattenbaum [74] reported on her application of neurofeedback training with a college age cellist, who suffered with daily migraine headache and disabling emotional distress. She applied guided imagery training and Open Focus™ exercises to enhance mental openness and receptivity, along with neurofeedback. Her client reported a remission of the headaches, won first chair in his conservatory, and graduated a year early.

Biofeedback and neurofeedback offer promising tools to assist children in their pursuit of artistic and athletic achievements. Emotional distress impedes excellence, and biofeedback can assist the child in alleviating distress. In addition, biofeedback can be used to increase resilience, enhance reaction times, and achieve optimal focus and attunement [68, 69].

Case Studies

The Case of Sally: Biofeedback for Panic Disorder

Sally was a 7-year-old second grader when she first visited our clinic. Sally suffered recurrent panic attacks that were most common at times of separation from her mother. The onset of Sally's anxiety began the previous summer when she became separated from her mother at a crowded carnival; an hour passed before the carnival security staff was able to reunite Sally and the mother. Sally spent that time trembling, with racing heart and profuse sweating, imagining that she would never see her mother again. Shortly thereafter Sally's mother had a mandatory work-related trip. Sally clung to her mother and cried for 2 h at the time of the mother's departure. After the mother's work-related trip, Sally began suffering anxiety when the mother left for work or attempted to leave her with the father while she ran errands. Both parents reassured Sally that they loved her very much and would not abandon her. She seemed to improve through the summer but suffered several anxiety attacks when school began, and separations became a daily event.

Over the first several weeks of school, Sally began to experience anxiety at other times, not related to separation. If she played hard and her heart rate increased, she began to fear that she was having another attack and that she might die. Her pediatrician tried a brief cognitive intervention, teaching Sally to recite that "I have a healthy heart, and I will be fine." Sally tried this coping strategy but broke down in tears when she could not slow her racing heart. Normal acceleration of the heart with exertion had become a recurrent cue for her panic.

Sally's parents were especially concerned because there was a history of anxiety disorders for at least three generations in Sally's mother's family. On her pediatrician's recommendation, they brought her for biofeedback-assisted psychotherapy for her anxiety.

Sally was frightened most by not being able to control the course of her anxiety attacks. Each time she noticed her heart racing and increased sweating, she suffered mentally with fears of never regaining control and "exploding" her heart. There was a family history of heart disease, and she recalled hearing that an aged aunt had "exploded her heart."

Sally's therapist provided further reassurance that children's hearts don't explode and that it is healthy for heart rate to increase with exercise. But Sally needed tools to manage her physiology, because her self-talk was not calming the severe autonomic activation, once she had begun to panic.

Sally had not specifically noticed hyperventilation, but her therapist observed both breath holding and a speeding up of breath when she discussed either separation or her fears of exploding her heart. It appeared that dysregulated breathing was driving some of Sally's anxiety symptoms. Sally's therapist used HRV and respiratory biofeedback to show Sally how her breathing controlled her heart rate. She could see on a computer monitor how her heart rate increased as she breathed in and slowed as she breathed out. The therapist used a "breath pacer" to help her vary her rate of breathing, first slowing it and feeling very relaxed and then hyperventilating moderately and experiencing some of her anxiety symptoms. Then, the therapist helped her slow her breathing again and self-regulate her heart. Sally was excited at having some initial control over her "out-of-control" body, and her therapist also taught her to begin saying to herself, "My heart is my friend, and I can make it pump faster and slower."

Sally mastered several new self-control skills during her weekly biofeedback sessions and in homework assisted by her mother. She mastered mindful diaphragmatic breathing, learned to warm her hands (aided by thermal biofeedback), and learned to use visualization to self-calm. One of her favorite visualizations involved using a kind of control dial, something that resembled a dimmer switch, to dial up and dial down her heart rate. Sally seemed to be a "natural" for self-regulation-oriented therapy. She loved the enhanced self-control. Her parents downloaded a breath pacer called EZ-Air Plus™ to aid her

home practice, and she made bets with her therapist about how much breath control she would show at her next session.

Within 3 weeks, Sally reported a cessation of anxiety attacks associated with elevated heart rate but still experienced some emotional loss of control and anxiety attacks when returning to school Monday morning. The therapist spent some time with Sally talking about normal separation fears and some of her past experiences of separating. Sally seemed to accept separation more and more on a cognitive level but continued to experience a racing heart and tearfulness on Monday morning when her mother dropped her off at school.

Sally wanted badly to extend her emotional and bodily control to moments of separation but felt stuck. Her therapist developed a plan with her and loaned her a digital thermometer and a StressEraser™, a portable HRV biofeedback device. In the car, on the way to school, she practiced warming her hands and diaphragmatic breathing, to create a calm state. Then, as her mother opened her door and hugged her goodbye, Sally "glued her eyes" to the StressEraser and created smooth sine waves of heart rate oscillation, maintaining a steady psychophysiological state as she hugged her mother.

The therapist also worked with Sally's mother Desiree, who had observed that her voice became shaky and she became tearful as she dropped Sally at school. Her emotions seemed to exacerbate Sally's emotional process. Desiree learned to use diaphragmatic breathing herself, during the drive to school, and also practiced looking at her watch and counting her breaths during the goodbye hug. This focus on their physiology served to dissociate both parties somewhat from the separation, while focusing them on physiological control.

One morning Sally's dad Bruce drove Sally and her mother to the school drop-off. Bruce got the giggles watching their ritual, as Sally stared at her StressEraser™ and strained to pace her breathing, and the mother stared at her watch and counted her breaths, while hugging goodbye. Fortunately the moment seemed perfect as Sally and Desiree were able to join in the laughter and appreciate the absurdity of the moment.

Sally made steady progress after the incident with her father's laughter. She was able to use paced diaphragmatic breathing in a less exaggerated form to calm herself at moments of separation and used self-talk to coach herself through separations. Sally completed 16 therapy sessions, including 12 sessions of respiratory and HRV biofeedback. She was also educated about the typical relapsing and remitting course of anxiety disorders and was assured that in spite of her family history of anxiety, her skills would aid her in any future episode of anxiety. Sally returned twice in the course of elementary school, each time requiring only two to three "refresher" sessions to reinforce her self-regulation skills and recover her self-confidence.

Questions

1. Which of the following factors contributed to Sally's anxiety disorder?
   (a) A traumatic separation from her mother triggered her initial anxiety episodes.
   (b) A family history of three generations of anxiety disorders suggests a genetic predisposition to anxiety.
   (c) Symptom generalization: The elevated heart rate accompanying initial anxiety episodes became a stimulus triggering cognitive fears and additional anxiety episodes.
   (d) Mental imagery of an "exploding heart," based on family history and dialogue, exacerbated Sally's fears.
   (e) All of the above.
2. Which of the following biofeedback techniques were *NOT* used in this case example?
   (a) Thermal biofeedback
   (b) Neurofeedback
   (c) Respiratory biofeedback
   (d) HRV biofeedback
3. Which of the following factors presented the greatest immediate barrier to recovery from her anxiety disorder?
   (a) Sally's breath holding and hyperventilation
   (b) Sally's separation anxiety and recurrent anxiety at routine separations from her mother
   (c) Sally's elevated heart rate during exercise
   (d) Sally's determination to regain control of her mind and body

Answers

1. (d): *All of the above*. Sally's family history showed three generations of women suffering anxiety disorder, with instances of panic attacks in each woman. Sally feared that this history condemned her to be anxious all her life. The history predisposed her to be vulnerable to anxiety, and her fears about the family history reinforced a helpless feeling. The separation at the carnival appeared to be sufficiently traumatic to sensitive her to normal everyday separations. The normal elevation of heart rate during activity later became a "conditioned stimulus" for panic, as Sally associated it with her anxiety attacks, triggering additional episodes. The story of her aunt's exploding heart and Sally's image of her own heart exploding served to reinforce her fear reaction to normal heart rate acceleration and escalate her cognitive anxieties. Each of these factors contributed in some fashion to the progression of her anxiety disorder and treatment needed to address each component in turn.
2. (b): *Neurofeedback*. Neurofeedback is frequently an effective tool in the treatment of anxiety disorders. Sally's treatment, however, emphasized (1) respiratory biofeedback and breath training, (2) HRV biofeedback and attention to voluntary heart rate control, and (3) thermal biofeedback training and general autonomic relaxation. When Sally warmed her hands, breathed in a paced diaphragmatic fashion, and produced large smooth heart rate oscillations, she felt much more subjective control of her body and her emotions. When she learned to voluntarily increase and decrease her heart rate through breathing, this gave her a tool for one of the most elusive aspects of her anxiety, the racing heart.
3. (b): *Separation anxiety*. Sally's determination to regain control of her mind and body were positive prognostic factors that made the self-regulation-oriented therapy more effective for her. She rapidly learned to pace her breathing and eliminate irregular respiration as a factor escalating her anxiety. She also learned to modify her heart rate voluntarily, through breath. So the elevated heart rate lost power as a stimulus for renewed anxiety. The separation anxiety remained the most difficult aspect of her anxiety to overcome.

The Case of Holly: Biofeedback Treatment of Headache

Holly was a 14-year-old, ninth-grade girl, referred by her pediatrician for migraine headaches that began following the onset of menstruation. Holly experienced only an occasional menstrual migraine at first, with one-sided throbbing pain, nausea, and sensitivity to smells and light. Over the course of 6 months, Holly's migraines became more frequent, persisting throughout menstruation and beginning to occur intermittently throughout the rest of her cycle. She also began to notice neck soreness, shoulder muscle tension, and muscle tension headache on a twice weekly basis, throughout the cycle.

On initial evaluation, Holly presented herself as a fairly well-adjusted happy girl, an honor student with high math ability; she reported perfectionistic strivings that contributed to academic success and positive experiences in school government but also contributed to some worry about course exams and academic abilities testing. Holly acknowledged growing anxiety and worry about headache but otherwise seemed emotionally well adjusted. Her initial biofeedback evaluation showed a baseline of elevated shoulder and neck tension and cool hands (83 F) and a pattern of tensing the trapezius (shoulder) and cervical (neck) muscles further, along with the frontalis muscles of the forehead, during a stress trial (mental mathematics and a visualization of difficult situations). Peripheral temperature also dropped more dramatically (76 F), and she engaged in some breath holding, during the stress trials. Holly did not recover well when asked to relax, after the stress trial. Her musculature remained tense and her hands remained in the 70s (F). She acknowledged a worry about causing herself a headache, and this worry seemed to contribute to some apprehensive tensing.

Holly's treatment plan called for a combination of in-office biofeedback training, including surface EMG biofeedback to enhance her muscle awareness and muscle relaxation, diaphragmatic breathing guided by respiratory biofeedback, and thermal biofeedback to master warming of her hands and fingers. Holly's therapist encouraged her to keep a journal in which she logged anxious and worrisome thoughts, times of muscle tightness or cool hands, and headache onset.

Holly learned progressive muscle relaxation and autogenic training in her treatment sessions and practiced these relaxation skills at home, guided by an educational CD. She herself came up with an additional suggestion, to enroll in a yoga class, which was available at the local YMCA. The Hatha yoga practices (Chap. 25), which included traditional yoga asanas (or postures) and an emphasis on yogic breathing, augmented what she was learning in her biofeedback sessions. Holly found herself much more aware of any onset of muscle tension and began to use her muscle relaxation and hand-warming skills, as well as positive self-talk, to manage these times of tension and was able to maintain a moderately relaxed upper body musculature and warmer hands most days, even during menstruation.

After 8 weeks of biofeedback training and psychotherapy, Holly reported a near cessation of the headaches outside the menstrual periods. The menstrual headaches were at first resistant to treatment, and her pediatrician considered hormonal intervention. However, neither Holly nor her mother wanted to begin hormone supplements, and this intervention was delayed. Holly began practicing yoga each morning at home and used the time on her school bus to practice diaphragmatic breathing and relaxation exercises. She also continued to journal anxious and worrisome thoughts, many related to academic perfectionism, and read a book called *Perfectionism: What's So Bad About Being Too Good* [75]. She succeeded in further moderating her muscle tension and worrisome feelings during menstruation and gradually experienced a lessening of the severity and frequency of her migraine headaches. After 4 months of weekly sessions, she decided to continue yoga classes and reduce the frequency of biofeedback training to monthly. By 1 year, Holly was able to report brief headaches occurring only one to two times per menstrual period on average and an occasional menstrual cycle without headache. Holly expressed pride at using a self-regulation approach and seemed to channel her perfectionism into a determination to continue her relaxation skills practices, her journaling, and yoga.

Questions

1. Holly's initial presentation included the following features which suggested a hormonal factor in her migraine headaches:
   (a) Holly's migraine headaches began after menstruation began, suggesting a hormonal trigger for her headaches.
   (b) Initially, Holly's headaches occurred only during menstruation.
   (c) The menstrual migraines persisted when all other headaches remitted.
   (d) All of the above.
2. Holly's biofeedback assessment showed the following features, suggesting specific biofeedback interventions:
   (a) Elevated muscle tension in the shoulders and neck and cool hands on baseline
   (b) Additional tensing of the shoulder, neck, and forehead musculature and further cooling of the hands during stress trials
   (c) Failure to relax the musculature and warm her hands during the period of directed relaxation
   (d) All of the above
3. Holly's comprehensive treatment did NOT include the following:
   (a) SEMG to facilitate relaxation of the musculature of the shoulders, neck, and forehead
   (b) Blood pressure biofeedback to moderate any arterial pressure contributing to her throbbing migraine pain
   (c) Thermal biofeedback to relax her autonomic nervous system and facilitate vasodilation
   (d) Yoga training and practice as an adjunct to biofeedback, to facilitate muscle awareness, muscle relaxation, and general relaxation
   (e) Cognitive behavioral therapy and bibliotherapy, to moderate perfectionism and worrisome thoughts

Answers

1. (d): *All of the above*. The onset of migraine symptoms after puberty commenced suggested hormonal etiology. The initial clustering of headaches during menstruation and the greater resistance of menstrual migraines to treatment all supported the conclusion that hormonal factors were a trigger for the overall headache problem and a factor contributing to chronicity.
2. (d): *All of the above*. Baseline indications of tense musculature and cool extremities are indications of a chronic stress response, present even in the absence of a specific stressor. The increase in muscle tension and the further cooling of the hands during a stress trial show that these physiological reactions were probably part of Holly's ongoing response to stress and were likely additional factors in the incidence and maintenance of headache. Her inability to reverse this stress response pattern by relaxation also showed that Holly continues to have an elevated stress response when the immediate stressful situation has ended.
3. (b): *Blood pressure biofeedback*. Arterial pressure does appear to be a factor in the pathophysiology of migraine. However, direct blood pressure biofeedback is a relatively ineffective training modality and was not used for Holly. Muscle biofeedback and thermal biofeedback are the most commonly used interventions for migraine and tension headache, and the greatest numbers of research studies show support for these interventions for headache. Yoga is a useful adjunct to biofeedback therapy, enhancing muscle awareness, muscle relaxation, and general relaxation. Cognitive behavioral therapy and bibliotherapy (reading about one's problem and possible solutions) are also frequently helpful for headache patients, including adults and children.

The Case of John: Neuromuscular Rehabilitation

A case published by Bolek [76] will be summarized here to illustrate the application of biofeedback-assisted neuromuscular reeducation. Bolek [76] reported on the biofeedback "habilitation" of a smile. "John," a 4-year-old child, was born with seventh cranial nerve palsy, affecting his ability to suck at birth and leaving a motor deficit affecting chewing on the left side and producing a facial droop. The smile was weak and included a left-side sagging of the lips, with other facial muscles overactive during efforts at smiling. The treatment objective was to habilitate John's smile. The term habilitation was used because John never possessed normal left-sided facial motor control; the objective was to establish adequate motor control for the first time.

A constellation of muscle training criteria were set up for John, including increasing activity of the zygomaticus muscle and an inhibition of the buccinator muscle. John sat on a bench in front of a video player, which ran the video forward each time he met the muscle training criteria. Improvements in the smile were recorded by still and video photography; his grandmother and teacher spontaneously praised his improvement in smiling without knowing of the biofeedback intervention. Biofeedback was delivered in 14 one-hour sessions, and John's improved smile was featured on the cover of the publication *Biofeedback: A Clinical Journal* in summer 2012.

Questions

1. Which of the following statements about the assessment of John's facial droop are true?
   (a) John's facial droop had a basis in seventh cranial nerve pathology, and therefore any behavioral intervention was inappropriate.
   (b) The presence of this pathology postpartum and its persistence through age four indicates the chronicity of this disorder, indicating a poor prognosis for treatment and suggesting that this intervention was medically inappropriate.
   (c) Some children with seventh cranial nerve disorders recover spontaneously, so this treatment, however successful, could not be justified until adulthood.
   (d) The positive improvement in John's smile shows that even when nerve damage is present, there is some plasticity to motor nerve

function, and function can sometimes be restored by neuromuscular reeducation.

2. John's biofeedback treatment consisted of the following:
   (a) Training the muscles throughout the face to relax
   (b) Training the muscles throughout the face to tense and increase motor nerve activation
   (c) Training one specific muscle—the zygomaticus—to increase in activation while at the same time decreasing activation in a second muscle, the buccinators
   (d) Alternately tensing all facial muscles and relaxing all facial muscles, to break up stereotypic responding
3. Challenges in conducting biofeedback with John included the following:
   (a) A relevant "reward" had to be created to reinforce John's working toward the specific treatment goals.
   (b) The therapist had to contend with the attention span of a 4-year-old.
   (c) The relevant muscles in a child this age are quite small and present measurement challenges.
   (d) The biofeedback training sessions rapidly fatigued John.
   (e) All of the above.

Answers

1. (d): *This case example shows* that even when a motor disorder is based on demonstrable nerve damage and even when it has a chronic course, biofeedback-based neuromuscular reeducation can often restore some motor function. As the original article made clear, 70 % of patients with idiopathic facial paralysis do recover, but the prognosis for spontaneous recover is greatly reduced if the paralysis persists more than 4 months. Chronic nerve-based disorders are resistant to treatment, but the psychosocial benefit to a child from an improved facial appearance is quite great and appears to justify the intervention. The clinical improvement of this child and several children in Bolek's reported clinical series shows that motor retraining using biofeedback has a reasonably positive prognosis.
2. (c): *Training one muscle to increase activation while a second muscle decreases in activation*. Simply relaxing or tensing all muscles in the face would not simulate normal muscle coordination, which involves a precise coordination of a constellation of muscles. Alternately tensing and relaxing would also be irrelevant. The zygomaticus elevates the lips and creates the smile, while the buccinators retracts the cheek toward the mandible, which strains the lips and disrupts the smile.
3. (d): *All of the above*. Each of the responses to this question presented a challenge to the biofeedback therapist. The therapist had to choose a reward to maximize John's efforts on the task and chose favorite videos which moved forward when John met the training criteria. The therapist designed a regimen of 2 min of training followed by 1 min of play, to cope with limited attention span. The therapist used custom-made miniature electrodes to measure accurately from these very small muscles. The biofeedback therapist, an occupational therapist, and the child's mother all remained present to entertain John and encourage his attention to the task. Training goals were also kept simple and limited to fatigue him less.

Conclusion

In summary, biofeedback offers an effective tool for pediatric clinical practice, alone or in combination with hypnosis in an integrative treatment program. Outcome studies show clinical efficacy for applications with anxiety, ADHD, chronic pain, diabetes, epilepsy, fecal disorders, headache, insomnia, and traumatic brain injury. Promising applications are also emerging for asthma, autistic spectrum disorders, functional abdominal pain, neuromuscular rehabilitation, and optimal performance. Case studies of biofeedback treatment interventions show the value for children, providing them with interventions

that emphasize self-regulation, voluntary control, and specific skill acquisition. As McGrady and Moss [77] have observed, achieving personal mastery and self-efficacy in health areas supports higher resilience and improves health outcomes. Biofeedback applications provide alternative treatment pathways for families preferring to rely less exclusively on pharmacotherapy. In some cases such as neuromuscular rehabilitation, biofeedback can offer hope when no medical treatment is available to alleviate a disorder. High-quality training and education in biofeedback and neurofeedback are available through professional associations, publications, conference workshops, webinars, and commercial training companies. The Biofeedback Certification International Alliance provides quality assurance by screening training programs and certifying those that meet the BCIA blueprints of skills and knowledge.

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Hypnosis for Treatment of Functional Symptoms in Children

21

Ran D. Anbar

Abstract

Hypnosis has been reported to be helpful in the management of many functional symptoms in children presenting to a medical practice including headaches, irritable bowel syndrome, and those arising as a result of anxiety. Therapy with hypnosis can be directed at the symptom, an underlying stressor, or both. Hypnosis can be taught to patients within several minutes and can empower them to deal effectively with the physical and psychological stressors that have contributed to their clinical presentation. Clinicians who learn to use hypnosis as part of their medical practice find that their ability to help patients has improved greatly. This chapter presents background, a brief review of the literature regarding hypnosis, and case studies. Also provided is a guide on how to use hypnosis clinically including links to video demonstrations.

Keywords

Anxiety • Asthma • Dyspnea • Headaches • Hypnosis • Irritable bowel syndrome

Background

Traditionally, western medicine has focused on the treatment of the body, with scant attention to the mind/body interactions that often play a large role in the development and persistence of disease. Failure to address patients' psychological states results in a therapeutic gap. This can be filled to a large extent by strengthening the clinician/patient relationship and further facilitated through the employment of hypnosis as a clinical tool. Clinicians who learn to use hypnosis as part of their medical practice will find that their ability to help patients has improved greatly. Hypnosis can be taught to patients within several minutes and can empower them to deal effectively with the physical and psychological stressors that have contributed to their clinical presentation [1]. Other tools to help address the

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mind/body gap include using cognitive behavioral therapy (Chap. 19), biofeedback (Chap. 20), and yoga (Chap. 25).

What Is Hypnosis?

Experts have not yet arrived at a consensus definition of hypnosis. It can be thought of as a state of focused attention during which an individual is more receptive to suggestions, be they delivered by a therapist (heterohypnosis) or by the individual himself or herself (self-hypnosis) [1, 2].

The clinical effectiveness of hypnosis is dependent on several factors including [3]:

1. Its application in appropriate clinical circumstances
2. The rapport established between the clinician and patient
3. The patient's motivation to receive help and apply hypnosis effectively
4. The clinician's hypnotic skills
5. The patient's natural responsiveness to hypnosis and willingness to practice this skill sufficiently

Applications of hypnosis to a medical practice are best learned by clinicians through attendance in experiential workshops sponsored or approved by professional societies such as the American Society of Clinical Hypnosis, the National Pediatric Hypnosis Training Institute, and the Society for Clinical and Experimental Hypnosis. Clinicians who have completed hypnosis training workshops learn to appreciate better the power of communication and usually find that their language and nonverbal interactions with patients improve, even when they are not employing formal hypnosis [2].

The latter part of this chapter provides a discussion and video demonstrations of hypnosis that can be used as a guide to using hypnosis clinically, but only after a clinician has received initial training. The use of hypnosis without adequate training at best is unlikely to yield optimal clinical benefit and at worst can be harmful. For example, the relationship between the clinician and patient may be weakened because of a hypnotic interaction involving inadvertent use of language that causes the patient emotional discomfort.

Brief Literature Review

Hypnosis has been reported to be helpful in the management of many functional symptoms in children. Therapy with hypnosis in a medical practice can be directed at the symptom, an underlying stressor, or both. For example, a child who presents with a complaint of shortness of breath in association with test anxiety could be taught how to use hypnosis as a calming technique, which would help decrease anxiety and its associated somatic symptoms. Alternatively, the reason(s) for the test anxiety might be identified through a discussion with the child or even through the use of hypnosis as a way to help the child generate insight. If a child identifies lack of sufficient preparation for tests as a reason for development of anxiety, the therapy can be directed at figuring out how the child could become better prepared. Perhaps the child has a low frustration tolerance. In this case, the child could be taught how to use hypnosis to focus better and remain calmer while doing homework, which would help the child become better prepared.

Failure to address a major underlying stressor in a patient with functional symptoms may prevent the efficacy of symptom-directed hypnosis. For example, a child who is bullied at school may develop irritable bowel syndrome, which provides the child a face-saving way of school avoidance. (Note the child may not be consciously aware of the connection between school and the abdominal discomfort.) Such a child is unlikely to improve with therapy directed at alleviating abdominal symptoms, because they provide important protection. In this situation, addressing the bullying is essential before therapy is directed at the somatic symptoms.

A major reason hypnosis is helpful in the treatment of functional symptoms in patients presenting to a medical practice is its benefit in the treatment of anxiety [4]. Psychosocial as well as physiologic stressors such as those caused by a disease can lead to development of anxiety and associated somatic symptoms [5, 6]. Children with chronic disease are especially prone to

develop anxiety related to their illness, with associated symptoms including headaches, dizziness, feeling faint, tachycardia, palpitations, chest pain, chest pressure, shortness of breath, muscle tension, tremors, paresthesia, gastrointestinal complaints, and insomnia [7]. A less common cause of functional symptoms in the medical setting is conversion of a psychological conflict into a symptom [7]. For example, a child who is scared about disclosing knowledge regarding a traumatic event may develop vocal cord dysfunction that does not allow normal speech. Thus, consideration should be given to introducing a large number of pediatric patients to therapy with hypnosis, which can ameliorate many somatic symptoms associated with anxiety or other psychological disorders.

Before hypnosis is applied in the treatment of functional symptoms, patients should undergo a thorough medical evaluation including history, physical examination, and appropriate medical tests, in order to consider conditions that would be better managed with other forms of therapy. For example, in one study, a quarter of children referred for hypnotherapy were found to have a condition for which they required medical evaluation and management [8]. Some of these children might have benefited from both medical management and instruction in self-hypnosis techniques. Thus, clinicians who can offer both therapies are well equipped to address a wider range of patients.

Functional symptoms that have been reported to respond to therapy with hypnosis span the spectrum of pediatric patients (Table 21.1) [9]. Some of the common applications of hypnosis in a medical practice are described below.

Table 21.1 Functional symptoms in pediatrics reported to respond to hypnosis

Functional disorders
Dysphagia
Dyspnea
Encopresis
Enuresis
Food aversion
Habit cough
Insomnia
Nail-biting
Needle phobia
Nightmares
Sleep terror
Sleepwalking
Thumb-sucking
Tics
Trichotillomania
Verbal dysfluency
Vocal cord dysfunction
Common conditions with frequent associated functional symptoms
Abdominal pain
Allergies
Asthma
Autism
Burns
Cancer
Cyclic vomiting
Cystic fibrosis
Diabetes
Eczema
Headaches
Hemophilia
Hives
Inflammatory bowel disease
Irritable bowel syndrome
Nausea
Juvenile rheumatoid arthritis
Pain (acute, chronic)
Reflex sympathetic dystrophy
Sickle-cell disease
Warts

Adapted from Gold et al. [9] with permission from John Wiley & Sons, Inc.

Asthma

Hypnosis has been reported as beneficial as an adjunctive therapy in the treatment of children with asthma in several reports [10–12]. Hypnotic methods that have been used successfully in the management of asthma include relaxation techniques, suggestions relating to symptom control and relief, ego-strengthening, and insight therapy. For example, in a 2-year randomized study of 28 children with asthma, those who were taught hypnosis had significantly reduced asthma severity and school absences [10].

Headache

If headaches in children are determined to be the result of tension or migraine, a substantial number of published reports demonstrate the efficacy of hypnotherapy in this setting [13–16]. For example, in an uncontrolled study of 144 children, the use of hypnosis was associated with a decrease of headache frequency from 4.5 to 1.4 per week ($p<0.01$), intensity decline from 10.3 to 4.7 on a 0–12 scale ($P<0.01$), and duration decline from 23.6 to 3.0 h ($p<0.01$) [15]. A quarter of these children reported resolution of their headaches after their self-hypnosis training.

Nocturnal Enuresis

Even if there is no significant initial emotional trigger of primary enuresis, by the time a child presents for evaluation, associated emotional problems frequently have developed. Prior to the application of hypnotherapy, a thorough medical evaluation should be completed including an assessment of physical abnormalities and a history of constipation or habits that can predispose a child to the development of enuresis, such as drinking caffeinated beverages before bedtime [2]. There have been multiple published accounts of successful use of hypnotherapy for children with enuresis [17–20]. For example, Kohen and Olness asked children to draw their concept of the brain/bladder connection. The children were then prompted to use hypnotic imagery to guide their brain and bladder to improve their interactions while they were asleep [2].

Recurrent Abdominal Pain

Multiple studies have described the benefit of hypnosis training for children with recurrent abdominal pain and irritable bowel syndrome [21–23]. For example, 53 children with these diagnoses were studied by randomly assigning them to treatment with six sessions of hypnotherapy vs. standard medical and supportive therapy. There was a greater reduction of pain in the hypnosis group. After a year of therapy, the use of hypnosis was deemed successful in 85 % of the hypnosis group as compared to 25 % in the other group [22].

Practical Application: How to Teach Hypnosis

Introducing Hypnosis

Prior to teaching patients how to use hypnosis, it is important to educate children and their parents/caregivers about the nature of hypnosis and to set appropriate expectations regarding the therapy [2, 24, 25]. Provision of the following information can help patients become more receptive to hypnosis:

1. *Hypnosis is a way to use your imagination to help yourself.*
2. *Hypnosis does not involve mind control. You are the only one who can control your mind.*
3. *Hypnosis is not sleep. You can use hypnosis to help yourself fall asleep, but when you are doing hypnosis, you can be aware of what is going on and stop at any time.*

The patients can be told that since hypnosis is in their control, it is their choice whether to employ it. Thus, their motivation is key. If the children are not interested or ambivalent about the use of hypnosis, typically no benefit is achieved from its use.

Patients and their parents should be offered an opportunity to share their perceptions of hypnosis, and any misconceptions should be addressed. Otherwise, such misconceptions may prevent the children or parents from being comfortable enough to allow successful use of hypnosis.

A clinician's expression of faith in a children's ability to help themselves can be very helpful in encouraging the children to allow hypnosis to be effective. The clinician might point out that the children already have mastered many skills in life, such as riding a bicycle or learning how to read, and therefore, this is proof of their ability to learn new skills.

As part of the interview before children are taught how to do hypnosis, the clinician should learn about the children's interests. Suggestions

about these interests can be incorporated into the children's hypnosis work. In this way, the suggestions are more likely to be relevant to the children than ones based on the clinician's notions of what might best capture the children's imagination.

Children's interest in hypnosis often is piqued by demonstrations of two aspects of hypnosis: words and imagery.

1. The power of words can be demonstrated through arm strength testing (Video 21.1). In this technique, the children are asked to extend their dominant arm to the side and instructed to resist the pressure to lower the arm that is exerted by the clinician pushing the arm down. (If the children are small, the clinician should push on the mid-arm rather than the wrist, in order to generate less torque.) The arm resistance exercise then is repeated while the patients are coached to say that they are "weak" or "strong." Children usually are amazed that their words affect their arm strength and thereafter are more apt to believe in the utility of hypnosis. The clinician then emphasizes that the children's words affected how his or her body reacted. If the children or observing parents express disbelief about the validity of the demonstrated effect, the children can be encouraged to carry out the same exercise with their parents.
2. The power of images can be demonstrated through some hypnotic induction techniques. For example, patients might be coached to imagine their hands as powerful magnets that attract each other and notice how their hands come together by themselves (Video 21.2). Or, patients can be directed to imagine holding a pail full of wet, heavy sand with an outstretched arm, while the other hand is holding strings to big helium balloons (Video 21.3). In this case, the arm that holds the pail typically falls, while the other arm frequently rises, seemingly outside of the patients' control. At that point, the clinician can emphasize that the children's imagery also can affect how their body reacts.

At the conclusion of this introduction to hypnosis, children usually are very open to the idea that hypnosis can help them better control their functional symptoms. If the children are not receptive to hypnosis at this point, the clinician should hold off on further hypnosis instruction because it is unlikely to yield an effective response. Instead, the clinician might need to consider whether sufficient rapport has been established with the patients or employ other therapeutic approaches.

Hypnotic Induction Techniques

In addition to the aforementioned magnets, sand pail, and balloons induction techniques, hypnosis also can be initiated in many other ways. For example, children might be coached to:

1. *Clasp your hands together and hold them over your head. Now, pull apart the index fingers as far as you can, and notice how, as your fingers come together on their own, your eyelids can become heavier and heavier* (Video 21.4).
2. *Stare at a spot above your eye level until your eyelids close on their own* (Videos 21.5 and 21.6).
3. *Roll your eyes back as far as you can, and imagine staring at a spot through the top of your head. Take a deep breath in, and as you let it out, notice how your eyelids can relax and close* (Video 21.7).

Hypnosis Intensification Techniques

Many children do not require achievement of hypnotic "intensification" (also referred to as "depth") in order to derive therapeutic benefit. While intensification may not be necessary, it can be useful to utilize such techniques as a way of helping children become more adept at and self-confident with self-regulation. Also, these techniques can be used to achieve hypnosis induction in some children.

Examples of deepening techniques include:

1. *As I count from 10 to 1, notice how you can go deeper and deeper into hypnosis with each number* (Videos 21.8 and 21.9).
2. After ascertaining that a patient likes riding in elevators, the following instruction can be provided: *As you imagine an elevator descend-*

ing, notice how you can go deeper into hypnosis every floor you go down (Video 21.10). (If the patient dislikes elevators, a suggestion to imagine riding in one will not be received well.)

3. *Imagine a favorite place that you find relaxing (or makes you happy.) This can be a place you have been to, would like to go to, or even an imaginary place. Notice what you might perceive with each of your senses there. What might you see? What might you hear? What might you smell? What might you feel if you touched it? If there is something to eat there, what does it taste like? Notice how the more you pay attention to your senses the more real the experience can become, and the more relaxed (or happy) you can become* (Videos 21.11 and 21.12). (Many pediatric patients have vivid imaginations and report feeling as if their imagined perceptions are real. Such patients can be queried "*What do you perceive?*" instead of "*What might you perceive?*")
4. Patients can be coached to relax muscle groups from toe to head, from head to toe, or from their stomach outwards (Videos 21.13 and 21.14). A variation of this technique involves suggesting that the patients tense their muscle groups before relaxing them.
5. Patients can be taught to engage in diaphragmatic breathing that involves allowing the abdomen to expand during inhalation while minimizing chest muscle movement (Videos 21.15 and 21.16). It can be suggested that the patients can calm themselves by inhaling slowly through the nose for a count of 4, holding their breaths for a count of 5, and exhaling slowly through the mouth for a count of 7. Repetition of such breathing cycles several times can be very relaxing.

Hypnotic Suggestions

Hypnotic suggestions that can be given to pediatric patients with functional symptoms can be directed at relief of anxiety, which underlies many of the symptoms. In my experience, anxiety relief alone, achieved through relaxation, is sufficient to achieve a significant improvement or resolution of the symptoms in approximately 70 % of referred patients to a pediatric medical practice. The remainder of the patients benefit from hypnotic suggestions related to their specific symptom or underlying psychosocial stressors. Examples of hypnotic suggestion include:

1. *Notice how relaxed you have become as a result of doing hypnosis* (Videos 21.17 and 21.18).
2. *Imagine your symptom as a color. Imagine a color that represents how you will feel when your symptom is better. Imagine the color changing from how it is now to how you would like it to be* (Video 21.19).
3. Hypnotic analgesia related to the functional symptom can be achieved using various methods including Magic Glove (Video 21.20), House of Control (Video 21.21), and Change of Sensation (Videos 21.19 and 21.22).
4. A posthypnotic suggestion that can be very useful in the treatment of anxiety involves rehearsing a gesture during hypnosis, which the patients associate with the feeling of relaxation (thereby "anchoring" it). Following a hypnosis session, patients are able to reactivate the feeling of relaxation achieved during hypnosis through the use of the relaxation sign (Videos 21.23 and 21.24). Alternatively, patients might be told, *Whenever you want to recall the feeling of relaxation you have achieved in hypnosis, take a deep breath* (Video 21.25).
5. Patients can be invited to give self-suggestions, which give them a feeling of mastery over the hypnotic experience (Videos 21.26, 21.27, and 21.28).
6. Imagined rehearsal of activities while in hypnosis can help patients cope better with the actual activities. For example, an athlete can improve his or her sports performance through mental rehearsal (Video 21.29).
7. Patients can utilize a state of hypnosis in order to better understand themselves, by reflecting on questions in a relaxed frame of mind. A technique that is helpful in this setting is to teach patients how to consider an issue from a different point of view (reframing), which

allows them to deal better with the issue (Video 21.30).
8. Techniques that help patients gain insight into their symptoms include use of ideomotor signaling (Video 21.31 [26]), automatic word processing (Video 21.32 [27]), and talking to an inner advisor (Videos 21.33 and 21.34 [26]).

How to End a Hypnosis Session (Trance Termination)

Complete trance termination is important so that patients return to their usual state of alertness and mental processing. Since most young children spontaneously go in and out of hypnosis as part of their imaginative play, ending a hypnosis session can be done informally with most children. However, formal completion of a hypnosis session often is indicated with older patients. Some signs of being in a hypnotic state are listed in Table 21.2. Clinicians should ensure that these signs have reversed by the time the hypnosis session is complete.

Examples of Formal Trance Termination

1. *As I count from 1 to 10, notice how you can become more and more alert, full of energy, and ready to face the day with each number I count* (Video 21.35).
2. The clinician can use a louder voice as the trance ends (Video 21.36).
3. Patients can be coached to attend to various environmental cues such as the feeling of their feet on the ground or their arms on the chair (Videos 21.37 and 21.38).

Table 21.2 Phenomena that can be associated with a state of hypnosis

• Dilated pupils
• Eyes defocus
• Eyelid fluttering
• Ocular tearing
• Facial muscles relax
• In adults, lack of gross body movements
• Jerky movements
• "Inability"/disinclination to talk
• Literalness
• Time lag in providing responses

An Example of Informal Trance Termination

Take all the time you need in the next few moments to complete your hypnosis experience. When the time is right, and you'll know exactly when that time is, just open your eyes and come back (Videos 21.39 and 21.40).

Validation

Following a hypnosis session, it is useful to help the patients understand that something has changed as a result of the hypnosis. This validates their ability to utilize hypnosis successfully. Such validation helps patients feel more confident in their hypnotic abilities and lays an important foundation for additional therapeutic success with hypnosis. Examples of suggestions that promote validation include:

1. *How do you feel differently now as compared to prior to the hypnosis? On a scale of 0 to 10, how relaxed did you feel before doing the hypnosis? How relaxed do you feel now* (Videos 21.41 and 21.42)?
2. Patients can be coached to use their relaxation gesture (described above, as hypnotic suggestion (4)). *Notice how you feel after making your relaxation sign* (Videos 21.42 and 21.43).
3. *Notice how your symptom changed following hypnosis* (Video 21.12).
4. *Did you learn anything surprising or helpful during hypnosis* (Videos 21.44 and 21.45)?

Younger Children

Hypnotic techniques need to be adapted for children whose developmental age is under 10.

Younger children tend to have fewer questions about hypnosis and therefore do not seek or need as much background information.

Hypnotic induction techniques are modified based on the age of the child. Preschool children can be guided through playing with toys or storytelling. School-age children can enter hypnosis through pretending to be a superhero, favorite television star, or even with use of magic.

Hypnotic suggestions with young children typically are related to the induction used. For example, it might be suggested that as a superhero does not have a particular symptom, when the child pretends to be this character, their functional discomfort will improve. Alternatively, a child might be instructed to tightly hold a magic pebble in order to activate it and achieve resultant relief from his or her symptom.

Caveats

Medical hypnosis should be conducted only by healthcare or mental healthcare providers who have been trained to utilize this technique. Individuals without healthcare training do not have the experience necessary to identify medical conditions that can occur in patients who have been referred for hypnotherapy. It is unlikely that direct harm might be caused by a healthcare provider who has not received sufficient training in hypnosis. However, indirect harm can be caused in a number of ways. First, as hypnosis is unlikely to work when used inexpertly, patients might conclude that hypnosis is not appropriate for their issues or that the issues cannot be modified by a psychological approach. This can be a harmful incorrect conclusion that may unnecessarily prolong the patients' discomfort or lead to additional ineffective treatments. Secondly, the healthcare provider might conclude that hypnosis is unhelpful in a particular clinical situation, which may be an erroneous assumption that will preclude other patients from receiving benefit from hypnosis [25].

This chapter has focused on the use of hypnosis in the medical setting. Patients who develop functional symptoms with an associated psychiatric disorder such as depression, bipolar disorder, or obsessive compulsive disorder should be treated by a mental healthcare provider. For such patients, treatment initially directed at symptom relief is unlikely to be fruitful given that the underlying psychosocial stressors have been not addressed. In such instances, the patients' symptoms may even worsen as a result of suggestions that they could be alleviated.

Case Studies

Case 1: Headache

An 11-year-old boy developed a constant headache 3 months before he was referred for evaluation with a therapist who uses hypnosis. The patient reported that his headache felt as if there was constant pressure involving his entire head. He explained that his headache intensity varied from a "6" to a "9" on a "0–10" scale, wherein a "0" represented no headache, while a "10" represented the worst headache he could imagine. He had noticed that his headache improved with rest and worsened when he was exposed to loud sounds or bright lights. The patient said he had no idea why his headache had occurred. As a result of his headache, the patient had been unable to focus at school and therefore had missed approximately half of the school days since onset of the headache in the late fall. The patient had been evaluated by his primary care physician and neurologist, but no abnormality was identified on physical examination, blood investigations, or cranial CT scan. Various oral medications were not of benefit.

The patient lived with his parents and 5-year-old sister. He was in 6th grade and said he liked school. He wanted to become a veterinarian. He enjoyed playing many different sports.

The patient was instructed on how to use hypnosis to help himself. He learned to utilize the magnetic hands and sand pail/balloons induction techniques (Videos 21.2 and 21.3) as a demonstration of his ability to change how his body reacts in response to his thoughts. He was asked to think of a calming activity and decided to imagine playing baseball. While utilizing this imagery, the patient was coached in progressive relaxation (Videos 21.13 and 21.14). He picked the anchoring gesture of making a fist as a reminder of how to trigger a relaxation response even when he is not in hypnosis (Videos 21.23, 21.24, and 21.25). When he was alerted, the patient reported that the baseball game appeared real and that he felt much more relaxed. He was excited to note that his headache had resolved for the first time in 3 months!

The patient was encouraged to continue to use hypnosis at home. However, the next day his headache recurred and did not improve with the use of hypnosis. Therefore, he returned for additional work with his therapist. It was suggested that the use of hypnosis appeared to be potentially useful for the patient since his headache had resolved transiently on the previous day. Furthermore, it was suggested that there may have been a psychological reason that the headache recurred and no longer was responsive to hypnosis. The patient agreed to allow his subconscious to express itself through typing while in hypnosis ([28], Video 21.32), at which point he disclosed that the headache had started because of his grief related to his dog's death 3 months beforehand. While in hypnosis, he expressed that he wanted to obtain a new dog and when he was alerted he became enthusiastic about discussing this possibility. Once again, the headache resolved following the hypnotic intervention and this time did not recur.

Questions

1. The use of hypnosis in treatment of a patient's headache would be appropriate in all of the following circumstances EXCEPT:
   (a) No physiologic cause of the headache has been identified after an extensive medical workup.
   (b) In order to verify whether a headache can resolve with suggestion, in which case it is unlikely to be related to an organic issue.
   (c) The headache has been attributed to migraines.
   (d) The headache has been attributed to tension.
   (e) The headache has improved with the use of amitriptyline.
2. This patient's headache resolved following the use of hypnosis without the use of imagery specifically related to the headache. Reasons for such improvement are likely to include all of the following EXCEPT:
   (a) The headache was related to anxiety, which was improved as a result of the hypnotic experience.
   (b) The patient realized that he would be able to change how his body reacted based on his experiences with the demonstrated induction techniques.
   (c) The patient believed that the hypnotic experience would be helpful to him.
   (d) When the patient thought about baseball, he focused his mind away from attending to the discomfort related to his headache.
   (e) The patient did not really have a headache, and the use of hypnosis provided him with a face-saving way of reporting that his headache had resolved.
3. The patient's development of insight regarding the cause of his headache likely was helpful to him because:
   (a) It provided him with an explanation of why the headache had occurred.
   (b) The typing method used to help derive the insight showed him that he was capable of remarkable hypnotic feats and thus gave him increased confidence in himself.
   (c) The extra attention during this process was key.
   (d) The acknowledgement of his wish for a new dog allowed for action to be taken to help him feel better.
   (e) None of the above.

Answers

1. (b): Hypnosis can be helpful in the achievement of headache relief regardless of its cause. Thus, the use of hypnosis may mask or attenuate a symptom that arises as a result of a physical illness that requires medical intervention, e.g., headache as a result of a brain tumor. Thus, hypnosis should only be offered for treatment of a headache after the patient's condition has been evaluated medically. Improvement of a headache with the use of medication does not preclude the use of hypnosis. Further, the use of hypnosis may help prepare the patient better for a trial of weaning a medication.
2. (e): The patient's beliefs about hypnosis are a key element of a successful hypnotic intervention. Thus, the patient's realization that he could help himself was important. The patient's development of rapport with his therapist allowed him to trust that hypnosis could help him, which is another key element.

Relief of anxiety through teaching patients how to redirect their thinking is one of the major ways that hypnosis can help. Patients falsely report symptoms on only rare occasions. A much more frequent occurrence is that clinicians fail to accept patients' report regarding their level of discomfort, which is harmful to the process of developing rapport.

3. (d): Occasionally, development of insight is key to resolution of a functional symptom because it allows for the underlying stressor to be reduced or eliminated. Knowledge of why a symptom occurred may not be sufficient to help a patient overcome it. While attention from a therapist or an increase of a patient's confidence can be therapeutic, these were more likely to be secondary benefits resulting from the insight development process. It should be noted that in many instances, development of insight is not necessary in the treatment of functional disorders [29].

Case 2: Irritable Bowel Syndrome

The patient was a 16-year-old who had experienced abdominal discomfort since the age of 3 years [30, 31]. At the time of his presentation for therapy with hypnosis, he reported nearly daily mid-abdominal pain, with associated diarrhea. Because of his gastrointestinal issues, he had a modified school schedule that started at 10:30 a.m., was unable to participate in sports as he would have liked, felt his social life was disrupted, and he was unable to travel easily. He had been diagnosed by a pediatric gastroenterologist as having irritable bowel syndrome when he was 7 years old. By the time he was 10 years old, he had undergone multiple diagnostic tests including radiological studies of his upper and lower bowels, an evaluation for gastroesophageal reflux, an electroencephalogram, and blood work, all of which were normal. His symptoms persisted despite dietary modifications and the use of multiple medications including aminosalicylic acid, atenolol, enemas, hyoscyamine, methylphenidate, omeprazole, prednisone, and diazepam.

The patient learned how to use hypnosis to help himself. He imagined going down an elevator that allowed him to relax with each floor it descended (Video 21.10). He imagined going to a concert by singer/songwriter Dave Matthews. He was instructed to imagine a bright healing light above his head. It was suggested that as it passed through his body, this light would heal every nerve, every fiber, every muscle, and every cell in his body. Following the hypnotic interaction, the patient reported the experience seemed very real and that the light felt warm and pleasant. He was instructed to utilize hypnosis on a daily basis in order to help himself feel better.

During the subsequent week, the patient reported that he had five pain-free days. Within 2 weeks, he had minimal abdominal complaints. Soon thereafter he returned to school with a regular schedule that started at 7:30 a.m. As he could now complete his school day earlier in time to participate in after-school athletics, he was able to become a member of the soccer team. He did not experience any significant recurrence of abdominal symptoms during the following 12 years.

Questions

1. At which point during this patient's clinical course could hypnosis been offered appropriately?
   (a) When he was 3 years old and just developed abdominal discomfort.
   (b) When he was diagnosed with irritable bowel syndrome at the age of 7.
   (c) After he had undergone all of the diagnostic tests at the age of 10.
   (d) After he failed all of the prescribed medical therapies at the age of 16.
   (e) Therapy with hypnosis for irritable bowel syndrome is not evidence based and therefore should not have been offered to this patient.
2. All of the following hypnotic suggestions might have had a similar effect to the healing light imagery EXCEPT:
   (a) *Imag*ine *a comfort dial that changes the quality of your discomfort when you turn the dial.*

(b) *Imagine a color that represents your abdominal discomfort. Now, imagine a color that represents how you want your abdomen to feel. In your mind's eye, turn your abdomen from its current color to how you want it to appear.*
(c) *Allow your hand to become warm as you imagine holding a warm drink at the concert. Once it is warm, apply the hand to your abdomen and let the warmth spread into your abdomen.*
(d) *Focus on sending deep thoughts to your abdomen and tell it to stop hurting and causing you to be sick.*
(e) *Imagine a peaceful inner self that spreads calmness throughout your body.*

3. What did the patient demonstrate when he reported that the hypnotic experience seemed "very real"?
   (a) The patient was a very good hypnotic subject.
   (b) He likely was saying so because he thought this would please his therapist.
   (c) The patient had unrealistic expectations of hypnosis.
   (d) The patient had a weak will.
   (e) The patient may have had mild psychosis as he could not tell the difference between reality and fantasy.

Answers

1. (b): There is an extensive medical literature supportive of hypnosis as an effective therapy for irritable bowel syndrome ([22], Chap. 5). Thus, this therapy could have been offered as soon as the diagnosis was made, when the patient was 7 years old. Had the therapy been successful at that time it would have obviated many of the diagnostic tests and trials of ineffective medication to which this patient had been subjected.
2. (d): All of the other suggestions involved the use of metaphors and focused on positive messages that suggested the patient will improve. Focusing on stopping pain involves a suggestion that the patient consider the experience of pain, which may intensify the discomfort.
3. (a): Report of a vivid hypnotic experience is one measure of a subject's ability to use hypnosis and thus makes it more likely that the use of hypnosis will be effective. When therapists are careful to be accepting of any patient response, it is less likely for a patient to report falsely regarding their experience. It is a myth that patients who are weak willed are better hypnotic subjects. This patient's report that his hypnotic experience seemed real does not mean that he cannot tell that the experience was imaginary.

Case 3: Complications of Prematurity

The patient was evaluated for increasing difficulties with verbal expression when he was 14 years old [32]. He explained that he had the most difficulty when he was at the center of attention, such as when his teacher called on him during class. Also, he exhibited a nasal tic (sniff or wiggle) for most of his life and seemed unaware of it.

The patient was the 600 g product of a 26-week gestation complicated by maternal autoimmune difficulties and preeclampsia. He was administered mechanical ventilation for 7 weeks and fed through a nasogastric tube for 12 weeks. His mother recalled that the patient batted at his nasogastric tube on many occasions, as he wrinkled his nose in apparent discomfort. He had a grade I intraventricular hemorrhage but did not develop neurological sequelae.

Upon the start of grade school, the patient sometimes was unable to express himself verbally because of difficulties with articulation. Initially, this was attributed to an oral motor problem, but the patient became visibly more frustrated with his speech pattern as he became older. At 9 years of age, he was diagnosed as having attention deficit disorder for which he was treated with methylphenidate. Apart from constitutional growth delay, he had no other health problems.

The patient was instructed in self-hypnosis calming techniques that he applied throughout the subsequent week. He found that he no longer experienced any major speech delays. At the subsequent visit, he was taught ideomotor signaling

(Video 21.31) through which the subconscious agreed to interact verbally (automatic talking). The subconscious expressed that the patient was sad about having been born prematurely. He explained that the patient disliked being subjected to teasing as a result of his small stature. After further discussion, the subconscious acknowledged that the patient had developed a wonderful personality as a result of learning how to deal with his medical difficulties. At the conclusion of the discussion, the subconscious stated that the patient now was happy about having been born prematurely.

A week later, the patient reported no difficulties with his speech and that his nasal tic had resolved. He pointed to a T-shirt he had picked to wear that day, which stated, "I have decided to put myself in charge."

Questions

1. What was the main indication that hypnosis likely would be helpful with this patient's speech issues?
   (a) Teenagers, such as this 14-year-old, tend to respond very well to hypnosis.
   (b) Speech problems resolve readily with hypnosis.
   (c) This patient's speech difficulty was exacerbated by social stress.
   (d) The patient had attention deficit disorder, which is a psychological issue.
   (e) The patient disliked having short stature.
2. Likely reasons that hypnosis helped resolve this patient's speech difficulties include all of the following EXCEPT:
   (a) The patient felt more confident in himself as a result of his success with the use of hypnosis.
   (b) The hypnotic calming techniques helped counteract anxiety that was underlying the patient's speech hesitancy.
   (c) The patient felt more in control of himself once he learned to employ hypnosis.
   (d) Hypnotic calming techniques helped improve blood perfusion into the patient's speech center and thus allowed him to speak more easily.
   (e) The knowledge that patient could regulate his body's reactions was empowering.
3. All of the following statements related to the resolution of the patient's nasal tic may be true EXCEPT:
   (a) The tic represented the patient's discomfort with having been born prematurely. Once his discomfort improved, his tic resolved.
   (b) The tic may have been related to the patient's anxiety, which improved following application of hypnosis calming techniques.
   (c) The tic may have been caused by memory imprinting in the patient's neonatal period related to his experience with the nasogastric tube.
   (d) The tic was related to the patient's reaction to socially stressful situations, which improved with the use of hypnosis.
   (e) The tic was completely under the patient's conscious control all along, but he did not realize it until he learned that he could regulate his own body better.

Answers

1. (c): Since the social stress appeared to exacerbate the patient's speech difficulties, it was likely that anxiety was related to at least some of his presentation. Hypnosis should be helpful in the treatment of such anxiety. While adolescents often are very open to the use of hypnosis, if they are not motivated, hypnosis is unlikely to yield much benefit. While some speech problems can improve with the use of hypnosis, it is an overstatement to imply that hypnosis can help resolve speech problems in general. The patient's diagnosis of attention deficit disorder and his issues related to his stature do not indicate that all of his symptoms would improve with a psychological approach.
2. (d): There is no evidence that hypnosis improves blood flow to the speech center. Furthermore, this patient's speech difficulty likely was related to anxiety, which is thought to involve a broad neuronal network including the limbic system, parieto-posterior cortex,

and cerebellum [33]. All of the other answers are likely.

3. (e): By definition, tics are not under complete conscious control (Chap. 2). Memory imprinting from infancy has been implicated in the persistence of symptoms in some cases [34]. In this case, the nasal symptoms may even have represented a conversion disorder, in that they resolved once the underlying psychological conflict was addressed.

Conclusions

Clinical hypnosis can help effectively and efficiently address psychological issues that usually underlie the development and persistence of functional symptoms in children. Hypnosis is best learned by clinicians through attendance in experiential workshops sponsored by professional societies.

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Guided Imagery for Functional Disorders

22

Moshe S. Torem

"Imagination is more important than knowledge."

(Albert Einstein)

"We are what we imagine ourselves to be."

(Kurt Vonnegut, Jr.)

Abstract

Therapeutic guided imagery (TGI) is a mind-body medical intervention that has the potential of providing healing benefits to individuals with functional disorders. Moreover, it is a skill that most patients can learn and practice on their own on a regular basis. TGI can promote healing, reduce suffering, and enhance the body's capacity to prevent and resist illness. TGI is a treatment modality that has no negative side effects and is based on experiencing a desirable positive outcome with one or more of the five senses (visual, auditory, tactile, olfactory, and gustatory).

Keywords

Guided imagery • Imagery • Meditation • Psychotherapy • Therapeutic imagery • Visualization

Background

This chapter describes the value and effectiveness of using guided imagery as a therapeutic tool in the treatment of children and adolescents with functional disorders.

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Guided imagery is defined by Astin [1] as involving the generation of different mental images evoked by one or more of the five senses (visual, auditory, tactile, olfactory, and gustatory). Such images are typically experienced with the goal of evoking a psychophysiological state of relaxation or with some specific outcome in mind (e.g., visualizing one's immune system attacking cancer cells, experiencing oneself feeling healthy and well, exploring subconscious issues, or imaging a solution to a current problem).

Bresler and Rossman, founders of the Academy of Guided Imagery [2], expand their definition of guided imagery to include experiences that are activated by a range of techniques that include simple visualization, meditation, prayer, artistic drawings, and storytelling.

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Guided imagery is differentiated from hypnosis by the fact that it does not require the patient to be in an altered state of consciousness or experience a state of dissociation. Other differences include that people who are in a hypnotic trance are more responsive to suggestions and frequently develop posthypnotic amnesia and their behaviors and physical sensations are often experienced as involuntary. These phenomena typically do not happen during guided imagery and are not necessary for therapeutic outcomes with its use. However, some patients may spontaneously shift into a trancelike state even if the therapist does not give any suggestions for the patient to do so. Clinicians should be aware of this phenomenon and verify that the patient has become fully reoriented to his or her immediate environment at the completion of a guided imagery intervention.

History

Imagery has its origin in many cultures dating back to ancient history. Shamans used imagery in their healing practices as mentioned by Achterberg [3]. In 1915, Carl Jung [4] recognized the value of imagery as an expressive vehicle for the subconscious mind. He then developed a psychotherapeutic method he called "active imagination" in order to help psychoanalytic exploration. In 1930s, the French psychotherapist Robert Desoille [5] described a treatment method he called "directed daydreaming." In 1989, a primary care physician Dr. Martin Rossman and the psychologist Dr. David Bresler cofounded the Academy for Guided Imagery, which began certifying health-care professionals in the use of guided imagery [2].

One of the common myths still lingering in psychiatry and medicine is that "what counts most is action and not fantasy." This myth was explained by Zilbergeld and Lazarus [6]. They both clarified that "how we picture ourselves and what we say to ourselves determines to a great extent how we project ourselves in the world." Human beings have historically believed in the power of the image to affect their reality. The traditions of using visualization rituals to heal humans from illness originated with the Shamans in primitive tribes and can be found through the practice of healing during the Greek era, ancient Chinese medicine, the practice of Yoga in India, the practice of Jewish Kabalistic healing rituals, prayer rituals and meditations in Christianity, and the tradition of the Sufis in the Muslim world. Scholars in the eighteenth century, which was proclaimed as the "age of reason," scoffed at any interventions utilizing imagery in the practice of medicine. As imagery phenomena could not be understood using a mechanistic model, the study and practice of imagery went underground. It became the province of secret, mystical, and para-religious societies.

The nineteenth century brought a willingness to openly accept the practice of imagery, meditation, and visualization, especially in association with the study of hypnosis, fantasies, dreams, and their effects on health and disease. Carl Jung in his book, Modern Man in Search of a Soul [4], said that "powerful images come to our awareness from all parts of our psyche." He called the center of our psyche the "self" or the "soul." He believed that this center produces images of great regulating power that have homeostatic effects for the mind and the body. Jung believed that mental images were, in fact, the most important reality human beings have.

In 1942, Jacobson [7] experimentally demonstrated that when we visualize ourselves running, the muscles of our legs, in fact, move involuntarily. This key experiment showed the link between the conscious mind and automatic responses in the nervous system. Jacobson went on to formulate his progressive muscle relaxation technique which relies on imagery and is utilized for stress reduction by health-care professionals. His technique continues to be used in present-day healing practices.

In 1951, Murray [8] developed the Thematic Apperception Test (TAT) that uses the interpretation of meaningful mental images as a diagnostic aid in the assessment of mental disorders.

By the mid-1950s, mental imagery was being studied and utilized by many disciplines. The Rorschach test was developed as a diagnostic

aid to help in the assessment of psychiatric disorders, based on the mind's tendency to create images triggered by random shapes such as ink blots.

In 1959, Schultz and Luthe [9] presented 2,400 case histories showing how their relaxation imagery was effective in helping patients recover from surgery, in the treatment of asthma, headaches, arthritis, back pain, diabetes, and other medical conditions. Their relaxation technique was called *autogenics*, and they emphasized images of heaviness and warmth in the limbs and nature-based images such as standing on top of a mountain, flying over the clouds, or watching a sunrise.

Assagioli [10] created a comprehensive theoretical framework for practice called *psychosynthesis.* In this system, he saw human beings as naturally gravitating toward having a harmony with their inner selves and the external world. Assagioli utilized imagery to aid in the diagnosis of emotional problems and facilitate their resolution.

In the 1970s, Cousins [11] promoted the use of imagery in the treatment of heart disease, hypertension, and chronic pain. Siegel [12] recommended the use of imagery in the treatment of cancer. Benson [13] demonstrated how use of imagery can activate the relaxation response in the treatment of essential hypertension and reduction of overall stress.

In 1983, Hall and Goldstein [14] showed that subjects could use their imagination to increase the number of circulating white blood cells and the blood levels of thymosin-alpha one hormone, which is utilized by the T-helper cells of our immune system.

Ader [15] and other researchers such as Pert [16] have focused on the emerging new science of psychoneuroimmunology, showing how thoughts, feelings, and mental imagery interact with our immune system.

Torem [17–21] reported on the value of utilizing imagery in the process of promoting healing and recovery from anxiety, depression, and trauma. Torem [22–24] described a special future-focused type of imagery designed to achieve a desirable treatment outcome for a variety of conditions such as hyperemesis gravidarum, depression, and autoimmune disorders.

Theoretical Framework and Rationale

Assagioli [10] proposed that the fundamental principle involved in the use of imagery is that every image has a motor tendency (defined as a tendency to initiate behavior through the activation of muscle movement) and every action requires a previous image for that action to be executed.

In 1976, Kroger, a physician, and Fezler, a psychologist [25], defined an image as a mental representation of our five sensory systems of sight, sound, touch, smell, and taste. They stressed the point that the enhanced experience of mental images can produce the same feelings and physiological responses as the actual objects and scenes we imagine. Kroger and Fezler also described the hypno-behavioral model that represents the combination of behavior therapy and hypnosis. According to Kroger and Fezler, the use of imagery deepens hypnotic states which, in turn, heighten the subject's feeling states and the imaginative capacities. "If all sensory inputs are stimulated there is a greater likelihood that autosuggestions and post-hypnotic suggestions will more readily be followed."

In 1987, two psychologists, Zilbergeld and Lazarus [6], distinguished between two types of imagery: separated and merged. In separated imagery, the subject is a spectator observing himself from outside the body at an imagined scene. It is like watching oneself on a movie or on a television screen. A person can scan the whole imagery scene back and forth. In merged imagery, a person's experience is from within his own body, and he cannot see what is going on behind him. In this type of imagery, the experience is much more intense and is similar to the experience of daily living, seeing the world through our own eyes, and experiencing the world with our own senses. According to Zilbergeld and Lazarus, this distinction is important because people react differently to these two types of imagery. Merged imagery is more intense and, therefore, harder to do. They recommend for beginners to start with the separated imagery first and only then, in certain cases, move to the use of merged imagery. In the treatment of phobias, it is important to start out with separated imagery where the individual

is encouraged to see things from a distance. Only when that is mastered is the subject then encouraged to use merged imagery.

Imagery has two elements in it: content and structure. Content deals with what is in the image, and structure deals with the clarity of the image, whether it is in color or black and white, the size of the objects in the image, the tone and the volume of the sounds, and whether the objects in the image are moving or still. Are the objects clear or fuzzy? Are the objects in the image you are observing close by or far away? The issue of structure has great importance since, at times, all that is needed is to change the structure of the image in order to make it more acceptable, thus giving the patient a sense of mastery over the image.

Zilbergeld and Lazarus [6] point out that if one wants to decrease the intensity of the subject's feelings regarding certain images, it is wise to use separated imagery and change the structure from clear into fuzzy, from color into black and white, from a large size into a small size, from loud sounds into low-volume sounds, and from being close by to being distant. On the other hand, if we want to evoke strong feelings in the patient and create activating power to these feelings, it is more effective to use merged imagery with high clarity, specific images, with color, with nearby life-size pictures, and in motion.

When using suggestions in combination with imagery, it is important to replace an old behavior with a new behavior [6]. For example, when an individual wants to stop smoking, it is important not only to help such a person experience himself/herself with guided imagery as not smoking anymore but also replacing the smoking behavior with something else such as chewing gum or sucking on a piece of candy instead of a cigarette. This is the case since it is very hard for people to imagine not doing something as a replacement for a previously unwanted action or behavior.

Practical Application

With a new patient, it is useful to start with imagery that is easy, pleasant, and comfortable for the patient. Consider introducing the use of guided imagery by putting it in the context of the patient's symptoms, complaints, or desire for change. For example, "Would you like to learn an exercise to reduce your stress and promote a general state of calmness and relaxation?" Most people respond affirmatively. Then ask the patient whether he/she is comfortable with a beach scene or pick another scene of their choice. The clinician should then proceed with the patient's choice of an image since he/she knows what best appeals to him/her. Many people choose an ocean beach scene, others may choose an inland lake, and some feel most comfortable in their own backyard.

Universal Imagery for Relaxation

The following is a sample script illustrating the use of mental imagery to produce a general state of calmness, comfort, and relaxation:

> *"Put yourself in a comfortable position, and let your hands rest on your lap. Let your eyelids close and keep them closed. Now, take a deep breath… hold it…that's right. Now, slowly exhale…let your eyes relax…and let your body float…keep on breathing comfortably like this…in and out…at your own pace (match your words 'in and out' with the pace of the patient's breathing). Now, as you keep on breathing like this…in and out…with each breath that you take this inner calmness is becoming stronger and spreading all the way from your head down to your toes…from top to bottom… inside out, and outside in."*

Then proceed to give the patient a suggestion regarding his/her ability and confidence in using such imagery in the future, and practice with them during the remainder of the session. For example, you may say:

> *"As you sit here feeling fully calm, relaxed, and buoyant, you need to know that you have obtained a new skill that is practiced in the future on your own. Anytime you want to experience this state of self-calming comfort with ease, all you need to do is put yourself in a comfortable and safe position, and count in your own mind from one to three. At one, you just look up with your eyes, all the way up to the top of your head. At the count of two, you slowly close your eyelids and take a deep breath. At the count of three, you exhale, let your eyes relax, and let your body float. The more you do this exercise, the easier and easier it becomes for you, and the more you do it, the more talented and experienced you become, and greater and deeper becomes the sense of total calmness and relaxation.*

Your mind, body, and spirit reach full harmony. This inner calmness and relaxation continue to create internal harmony within you, putting in synchrony various systems and organs in your body, working harmoniously together to promote your healing and recovery. This calmness and relaxation continues to stay with you as long as you need. The way to come out of this state of imagery is to count back from three to one. At the count of three, you get ready to shift to a regular state of attention, at the count of two with your eyelids closed, you look up with the eyes, and at one, the eyelids open, and your gaze comes back into focus. You are becoming fully alert and awake, oriented to your surroundings, ready to deal with your tasks of daily living adaptively and effectively. That's right. Let's go ahead and do it together right now. Three, get ready in your own mind. Two, with your eyelids closed, look up with the eyes. That's right, and one, the eyelids open, the eyes come back into focus. You are becoming fully alert and awake, and this inner state of calmness continues to stay with you as long as you need it. Good. How are you feeling right now?"

Most patients report a pleasant experience of calmness and relaxation. Some of them even say spontaneously, "I have never felt so relaxed in my whole life," or "This is great! I love it! You mean I can do it on my own?" then proceed by saying the following:

"It's impressive how well you have learned to experience your imagery. In fact, you need to know that you can do this on your own in the future, since you have had this gift for self-evoked imagery all of your life. Today, you have learned how to evoke this state of mind on your own. Now, I would like to do this exercise with you again. I serve as your coach, and you are my student-athlete. Are you ready?"

Then go through the exercise again, this time being less wordy, allowing the patient to use his/her own cues to count themselves into and out of self-evoked imagery, and if necessary, repeat it a third time. Patients are instructed to practice this exercise on their own three to four times a day in a safe environment. In the following session, spend some time discussing the patient's practice of self-evoked imagery, and allow time for clarifications and minor "tune-ups" (for a verbatim example, please refer to [21], pp. 239–241).

Most patients respond very well to this form of guided imagery and it can be tailored to meet the patient's choice of images. In patients who also have insomnia, teach them to use this method to induce natural sleep by instructing them to practice it when they are in bed. These patients can be instructed to continue to focus on their nature scene or special comfortable place and given the suggestion that the next event they will remember is waking up in the morning refreshed, alert, and full of energy. This occurs since during the night they automatically shift from self-directed imagery into natural sleep.

Symbolic Guided Imagery

In one version of this method, the patient is guided into a state of mind inducing relaxation and calmness in a nature scene of the patient's choice. This is followed by the use of symbolic guided imagery intended to introduce a variety of natural images communicating changes of maturation, differentiation, integration, growth, self-mastery, control, and/or freedom of choice.

For example, clinicians can use images for gaining a sense of control and mastery by asking patients to experience themselves driving a car or riding a bicycle, controlling the steering with both hands, changing the speed of travel, changing lanes, and using the brakes and other control instruments. All of these are suggested in association with a sense of pleasure and self-mastery.

Another image to use is one of the patients remodeling and redecorating their room or their home. The room is analogous to the patient's body. First, the patient can imagine living in an old room with which he or she feels dissatisfied. Then, the patient is coached to use imagery in which the room is remodeled and redecorated to meet individual needs. Emphasis is put on the patient's choice of colors, materials, furniture, drapes, pictures, etc.

Another effective image is that of the patient adopting a puppy or kitten, perhaps a sick one from the animal shelter of their local community. Then, the patient can be instructed to imagine the kitten or puppy nursed into full health through the patient's commitment, perseverance, and dedication in caring for the neglected pet. The sick pet is a metaphor for the patient's

unhealthy body for which they make a commitment to help and nurse back to health.

"Back from the Future" Technique [22]

Utilizing this technique requires understanding and familiarity with the patient's condition and life circumstances. A discussion is held with the patient about a desired future image in which the patient would be comfortable as representing a better and healthier life. Once that is identified, the patient is guided into a state of calmness. This is followed by suggestions for imaginary time travel into a specific time in the future. The future reality is enhanced by suggestions focused on visual, auditory, touch, smell, and taste senses. In addition, the experiences are enhanced by suggestions of positive thinking, pleasant feelings of joy, and pride in reaching a solution to a specific problem. This is accompanied by suggestions for a sense of health, strength, accomplishment, and a sense of inner resourcefulness and creativity in coping with life's stresses. Patients are then instructed to store the above positive feelings, images, and sense of accomplishment and take it with them as they are guided back from the future into the present. They are told that these positive images, sensations, and feelings are a special gift that they bring with them on their trip "back from the future" into the present and that these gifts will guide them on a conscious and subconscious level on their journey of healing and recovery.

When the patient is back to his/her regular state of mind, a brief discussion is conducted about the patient's experience. This is followed by a homework assignment in which the patient is asked to write down the experience and what it was like to take such a voyage into the future. The patient is asked to bring to the following session the written assignment. What happens frequently when using this technique is that if the patient uses the past tense in the written assignment, it is an excellent prognostic sign that this technique has worked and that the patient has internalized the suggestions and experiences from his "trip" into the future. Many of the symptoms of futurelessness (feeling as if there is no future), helplessness, and hopelessness are significantly reduced and are replaced by a sense of new hope, strength, and belief in one's own recovery. This technique has been successfully used in patients with eating disorders [21].

Case Studies

Case 1: A Boy with Nocturnal Enuresis

Ryan, a 12-year-old boy, was brought to the office by his mother for the treatment of nocturnal bed-wetting. He was fully worked up by his pediatrician and a urologist, and all organic diseases were ruled out as a cause for his continuing night bed-wetting. The use of imipramine produced a partial response, but then it was discontinued due to side effects. Ryan was found to be a very imaginative boy. He was described by his teachers and parents as being a "daydreamer." Frequently Ryan became so involved in his computer games or in reading a book that his mother had to come into his room and gently touch him to get his response to her request to join the family for dinner, since he did not respond to verbal requests communicated from another room.

He chose the ocean beach scene as his favorite vacation place. With guided imagery, he experienced the ocean beach with his five senses (visual, auditory, tactile, olfactory, and gustatory). The use of interactive guided imagery allowed him to describe in detail his experience of walking on the beach searching for seashells and collecting them into his bucket. When instructed to come back from the ocean beach, he reported with a smile how calming and relaxing it was. He was instructed to practice this exercise on his own at home. His mother was informed of it and agreed with the plan.

In the second visit, the discussion was about his dreams for the future. He stated that in his daydreams, he experiences himself being a doctor who takes care of sick children and how enjoyable this work is for him. When asked if he would be willing to fly an imaginary time

machine into the future and then experience himself as a mature, adult, and practicing pediatrician, he responded affirmatively. Future-focused guided imagery was used and was combined with positive suggestions. Ryan spoke of himself as a practicing doctor and a mature adult who obtained knowledge and wisdom; he was asked how well he is in control of his bladder. He reported having full control, night and day. He was then asked to deposit these experiences in the memory network of his brain and let his brain activate the proper controls of his bladder. He was given the suggestion to reexperience his imagery of the future with all his five senses and his feelings and sense of mastery that he attained over the problem he used to have as a child. He was also given the suggestion to bring back the skills, the knowledge, and the wisdom his body attained and let these experiences be special gifts that he brings back from the future as he returns with his imaginary time machine to the present. He was given suggestions that these experiences and body wisdom are now encoded consciously and unconsciously so that this mature wisdom from the future has been downloaded into the "hard drive" of his brain and is available to him anytime, day and night whether he is consciously aware or not aware or if he is awake or asleep. When asked to open his eyes, he smiled and reported that he liked his trip into the future stating that he was treating kids who have a problem with night bedwetting and had been curing them of that problem.

Two weeks later, he came for a follow-up visit and reported 100 % success of dry nights. This was confirmed by his mother. A follow-up visit 3 months later confirmed continued success.

Questions

1. What kind of imagery was utilized in Ryan's treatment?
   (a) Separated imagery
   (b) Explorative imagery to find the cause of his problem
   (c) Merged imagery
   (d) Interactive imagery
   (e) c and d
2. What was the mother's role in the treatment of Ryan?
   (a) The mother was present in all 3 sessions.
   (b) The mother was informed of the treatment plan.
   (c) The mother gave her consent to proceed with the treatment plan.
   (d) The mother was asked to report her observations of Ryan's progress.
   (e) b, c, and d.
3. What is the significance of Ryan's natural future-oriented daydreams?
   (a) Most kids of Ryan's age are present focused and do not daydream about the future.
   (b) Daydreaming about the future is common in boys of Ryan's age.
   (c) Experiencing the beach scene as a first step in learning imagery is helpful.
   (d) b and c.
   (e) None of the above.

Answers

1. (e): Most cases of nocturnal enuresis are understood as a functional symptom in children and adolescents. The use of merged imagery to enhance the experience of calmness, serenity, and other positive states of mind is more effective than the use of separated imagery. Enhancing the patient's participation in the process and improving the chances of desired effects of suggestion are accomplished with interactive guided imagery whereby the patient reports on his experiences during the imagery intervention as was done in this case.
2. (e): It is important to remember that the successful treatment of children and adolescents frequently needs to involve the parents. The patient's parents can potentially sabotage any well-intentioned interventions. Therefore, the patient's parent has to not only be informed of the treatment plan but also give their consent and acceptance of such a plan. Moreover, the patient's parent is given a particular assignment, in this case making observations and to be prepared to report them to the doctor in future sessions, as was done in this case.

3. (d): To enhance the successful use of guided imagery, it is important to provide the patient with a first experience that is calming, comfortable, and easy to use—as was done in this case. This first experience becomes the foundation for additional more sophisticated guided imagery interventions. Children of Ryan's age commonly daydream about the future whereby they see themselves as adults who are able to exercise greater freedom of choices and actions with more mastery and control of their personal lives. Knowledge of this natural phenomenon is important since it can be utilized in designing guided imagery interventions focused on a future desirable outcome of treatment.

Case 2: A Girl with Irritable Bowel Syndrome

Jennifer, a 17-year-old high school girl, was referred by her pediatrician for the treatment of irritable bowel syndrome (IBS). Her major symptoms included abdominal pain associated with gas and irregular bowel movements fluctuating between constipation and diarrhea. She was examined by a gastroenterologist who told Jennifer and her mother that she suffered from IBS. Jennifer was an excellent student performing academically in the top 5 % of her class. She was very meticulous about her work and was obsessed with cleanliness and organization of her room, personal items, her books, and homework. She would sometimes spend up to 30 min or more in the bathroom attempting to cleanse her bowels of what she believed was still containing undesirable stools. In the first session, Jennifer reported that when she goes on vacation with her family during the summer to their lake resort rented cottage, she does not think about this issue and enjoys row boating and hiking in the woods. During those times, her IBS symptoms are significantly alleviated.

Jennifer was asked if she wanted to learn how to take herself, with the use of imagery, to her favorite spot on the lake. There, she likes to hike and sit on top of a large boulder that allows her to view the scenery of the lake and the woods. She responded affirmatively, and with the use of interactive guided imagery, she reported in detail walking on her familiar trail in the woods toward her favorite spot. She experienced her walk visually, auditorily, and with her sense of touch and smell. As she was sitting on her favorite boulder, she was able to experience a state of complete peace and serenity. Ego-strengthening suggestions were given with such statements as follows:

> *"Now that you have reached your favorite spot, experience this place with all of your five senses. Notice the inner state of peace and serenity that is created with the beauty of nature surrounding you. Notice how your mind and body are now working together in harmony resonating with each other for the sake of Jennifer as a whole person. Internalize this state on a conscious and subconscious level. Download it to the "hard drive" of your brain and let it be available to you in the future as a healing tool. Let these experiences be encoded in your conscious and subconscious mind and even if you do not fully remember every detail of your experience, your subconscious mind will know and your body will match this state of mind in calmness, tranquility and in harmony with the natural needs and functioning of your body including all of your intestines."*

She was instructed to practice this exercise at home on a regular basis at least twice a day. At a follow-up visit 4 weeks later, Jennifer reported a significant improvement in her IBS symptoms. She brought in her personal diary and stated that she has been expressing her frustrations by writing in the journal and then practicing her visit to the cottage on the lake thereby internalizing the peace and serenity she had achieved from such a visit. She agreed to read from her diary and selected specific entries that illustrated her new skill of using words to express her feelings and emotions combined with her guided imagery trips to the cottage on the lake. A follow-up visit 2 months later confirmed her continued improvement in her IBS symptoms.

Questions

1. Who should choose the favorite spot for calmness and joy?
   (a) The patient's treating clinician
   (b) The patient's parent

(c) The patient's primary care physician
(d) Jointly by the patient and treating clinician
(e) Any of the above

2. In this case, the patient experienced her favorite spot with only four senses (visual, auditory, tactile, and olfactory). Which of the following statements about this patient is most likely to be true?
   (a) The chances of a successful outcome are not related to the patient's favorable response in the office.
   (b) The chances of a successful outcome are not related to the patient's continuing to practice the new skill at home on a regular basis as instructed.
   (c) Experiencing the imagery with only 4 senses does not reduce the chances of success.
   (d) The patient's chances of success are not influenced by members of the family and their attitude toward imagery as a treatment modality.
   (e) The patient's chances of success with therapeutic imagery are not affected by the attitude of her primary care physician.
3. How does therapeutic guided imagery (TGI) affect the gastrointestinal system?
   (a) TGI mediates brain centers that reach the GI system through the sympathetic and parasympathetic autonomic nervous system.
   (b) TGI contains a psychophysiological component.
   (c) TGI may reach the GI system by other yet unknown ways.
   (d) a, b, and c.
   (e) None of the above.

Answers

1. (d): A common reason for unsuccessful outcome in the use of TGI is when the clinician takes charge of deciding what specific spot will be associated with sensations of calmness and tranquility for the patient without the patient's participation in this process. Inviting the patient's participation in such a decision enhances the therapeutic alliance and the chances for a positive outcome.
2. (c): Experiencing TGI with only four senses can be very effective for many patients. It is important to encourage the patient to practice guided imagery at home and to get the patient's family and the primary care physician on board with the treatment program in order to enhance the chances for a favorable outcome.
3. (d): TGI activates brain centers and neural pathways involving the autonomic nervous system (sympathetic and parasympathetic). The pathway occurs through the vagus nerve that reaches the celiac plexus and from there the whole gastrointestinal system. This allows mediation of gastrointestinal motility and secretion of mucus, digestive enzymes, and other functions such as the reabsorption of water content in the large intestine [26].

Case 3: A Girl with Tension Headaches

Jana, a 16-year-old high school girl, was referred by her pediatrician for the treatment of her tension headaches with the use of guided imagery. Her symptoms were described as starting with a diffuse and dull ache in the back of her neck, then spreading to the back of her head, and then to the whole head. She explained she felt as if there was a round vice surrounding her head with pressured dull and constant pain. She had numerous tests and x-rays including a cranial CAT scan and MRI that all were normal. The use of ibuprofen and acetaminophen were only partially helpful by making the pain duller but not alleviating it completely.

When asked to provide any additional information about the headaches, she stated that they started occurring when she was still in elementary school. At that time, while riding on the bus, she was bullied by other girls and felt scared and intimidated because she was unable to stand up for herself and did not have any allies to protect her. When she eventually reported what happened to her parents, they told her she has to learn to stand up for herself. She described those bullies as "a pain in the neck."

In high school, when she faced similar situations, she felt alone and isolated; she developed severe headaches and would be allowed to leave

school and stay home for a couple of days until she got better. She stated that when she prayed and asked for God's help and protection to better deal with her predicament in school, she would feel better and the pain subsided.

When asked for her ideal imagery spot, she described a ritual of cleansing herself in the holy waters of the Jordan River located in the Holy Land. The guided imagery exercise in the office allowed her to experience this special site in the Holy Land. She reported feeling cleansed and strengthened, empowered to stand up for herself, and feeling now worthy of protection by God's special angels, Gabriel and Raphael, who will always be with her to keep her safe and protected. She was instructed to practice this exercise at home and then write an essay about it and her experiences. She was asked to bring the essay with her to the follow-up session.

At the following visit, review of her written essay revealed that she described her experience in the Jordan River by using the past tense. She then went on to describe additional writings she has put into her personal diary. In one of these, she describes a new experience of friendship in real life with two boys who were new to the school system and whom she first met at the bus stop. They were both kind and found the information she gave them about the school system and the teachers as very helpful. This started a friendship whereby these two boys sat with her on the bus and gave her a sense of safety and confidence. She reported a significant reduction in the intensity and frequency of her headaches.

At her follow-up visit 2 months later, she reported sustaining success in being free of the tension headaches while practicing her therapeutic imagery. She had deepened her friendship with the two new students and had made another new friend as well.

Questions

1. What type of guided imagery was used in the treatment of Jana?
   (a) Direct suggestions to alleviate the pain
   (b) Direct suggestions to alleviate muscle tension
   (c) Indirect suggestions through cleansing, calming imagery of immersing her body in the waters of the Jordan River
   (d) Suggestions that God will heal her predicament
   (e) Suggestion that new medications will heal her headaches
2. What was the role of her writing assignment in her treatment?
   (a) The writing assignment reinforced the power of her experience of practicing at home.
   (b) The writing assignment served as feedback to the doctor as to whether this imagery has been internalized.
   (c) The writing assignment provides feedback as to whether the patient has practiced the exercise.
   (d) The writing assignment allows for reporting new experiences.
   (e) All of the above.
3. Experiencing a solution to her problem in a future-focused imagery setting is of therapeutic value due to the following reasons:
   (a) It allows for experiencing a solution in a safe environment.
   (b) It allows the patient to have a corrective emotional experience in the session.
   (c) It works in part due to internalizing positive suggestions.
   (d) It activates the mechanism of a self-fulfilling prophecy.
   (e) All of the above.

Answers

1. (c): This case illustrates the importance of inviting the patient to become an active participant in choosing the proper imagery that will be most effective in reaching the desired therapeutic outcome. It also shows that an effective treatment plan is a co-creation of both the clinician and the patient. The indirect suggestions for cleansing and calming imagery were enhanced with ego-strengthening suggestions for achieving the best therapeutic outcome, which helped improve the patient's self-esteem and confidence. Her personal religious faith was respected in allowing her to choose her own imagery.

2. (e): Writing assignments reinforce the patient's experience of practicing guided imagery at home and also as a tool that provides the clinician with feedback regarding the efficacy of the therapy in the patient's natural environment. Moreover, the language used by the patient in the writing assignment provides the clinician with metaphors and phrases that are part of the patient's imagery and vocabulary. These can later be utilized by the clinician in designing suggestions and wording that is most compatible with the patient.
3. (e): The use of future-focused imagery is a powerful and safe intervention that enhances the chances of achieving the most desirable outcome of treatment [22, 24]. In addition, patients experience a solution to their problems during the sessions; they have corrective emotional experiences that can serve as a positive foundation for successful future interventions and experiences. The internalization of future-focused therapeutic experiences reinforces the pathways of success by activating the natural mechanisms involved in a self-fulfilling prophecy as described by P. Watzlawick [27].

Conclusions

TGI can be utilized as a powerful treatment tool in the practice of treatment interventions. However, it has been rather underutilized in the practice of Western medicine. The specific case reports and techniques described in this chapter serve as examples to motivate the adoption of using imagery more frequently in the treatment of functional symptoms.

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Acupuncture for the Treatment of Functional Disorders in Children

23

Anjana Kundu and Rosalie F. Tassone

Abstract

Increasing numbers of patients in the USA, both adults and children, are pursuing complementary and alternative therapies for health care. Acupuncture is one such therapy, which has been utilized in health care for thousands of years in China and other Asian cultures. This chapter provides a brief background about acupuncture therapy, various modalities that comprise the practice of acupuncture, and evidence related to its use with pediatric patients and functional disorders. We also present case-based scenarios to demonstrate successful use of acupuncture therapy.

Keywords

Acupuncture • Acupressure • Moxibustion • Cupping • Qi • Functional disorders • Constipation • Abdominal pain • Headache • Licensed acupuncturist • Needle phobia • Medical acupuncturists

Background

Acupuncture

Acupuncture is an ancient treatment modality developed in China over 3,000 years ago. The term *acupuncture* is derived from the Latin words *acus* (meaning needle) and *pungere* (meaning pricking). The National Center for Complementary and Alternative Medicine (NCCAM) defines acupuncture as "a family of procedures involving the stimulation of points on the body using a variety of techniques. The acupuncture technique that has been most often studied scientifically involves penetrating the skin with thin, solid, metallic needles that are manipulated by hand (Fig. 23.1a, b) or by electrical stimulation (Fig. 23.2). Practiced in China and other Asian countries for thousands of years, acupuncture is one of the key components of traditional Chinese medicine" [1]. The "acupuncture points" are located throughout the body along a network of channels called "meridians." Besides manual needle and electrical stimulation (electroacupuncture), acupuncture

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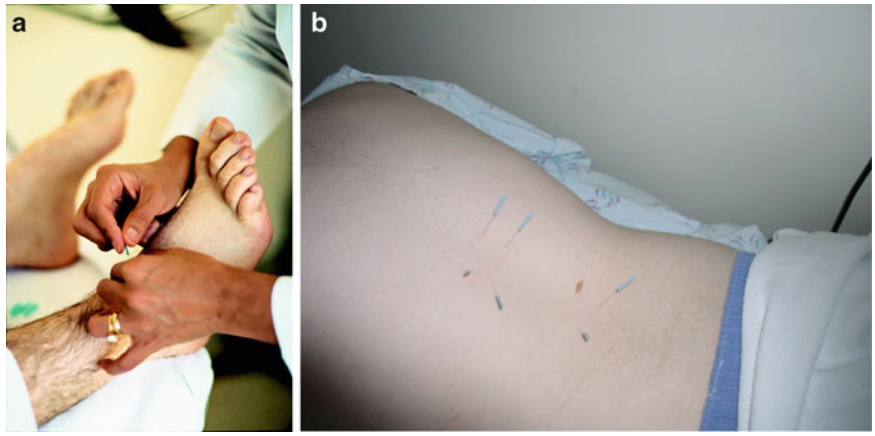

Fig. 23.1 (**a**) Acupuncture treatment with manual needle stimulation. (**b**) Acupuncture treatment with manual needle stimulation

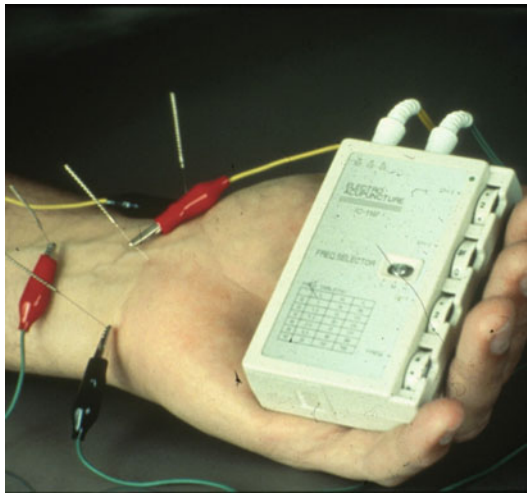

Fig. 23.2 An acupuncture treatment using electroacupuncture

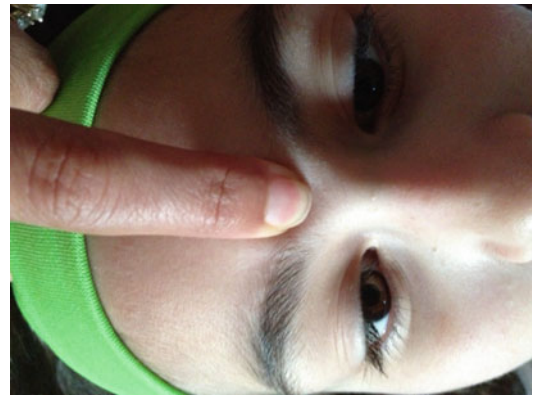

Fig. 23.3 Acupressure at Governing Vessel (GV) 24.5 or Yintang

points can also be stimulated using manual or digital pressure (acupressure) (Fig. 23.3), heated herbs (moxibustion), cupping (using negative pressure or suction via glass cups), laser (low-intensity IR lasers beam), or magnets (Fig. 23.4).

Conceptually, acupuncture is a technique used to promote and restore an unobstructed flow of *qi* (pronounced "chee") in the body. *Qi* has been described as a vital energy or life force that is believed to circulate in our bodies along specifically described acupuncture meridians ensuring harmonious function throughout our body,

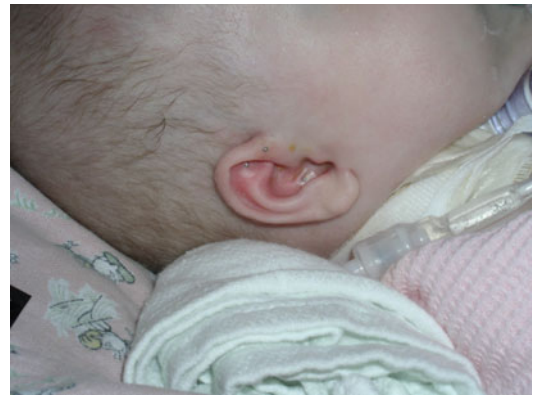

Fig. 23.4 An infant receiving acupuncture treatment with magnets at ear points

physiologically referred to as a state of homeostasis. Obstruction in the flow of *qi may* progress to an illness. A state of health can be restored by manipulating specific acupuncture points to allow for unobstructed flow of *qi* along the meridians.

Evidence for efficacy of acupuncture has been demonstrated in both basic science and clinical studies. Several basic science studies have demonstrated an increase in endogenous peptides such as endorphins, enkephalins, and dynorphins [2–5]. Non-analgesic effects of acupuncture have been shown to be mediated through its anti-inflammatory or immune-modulating effects [6, 7]. In addition, expression of serotonin in response to acupuncture stimulation has also been implicated as a mode of action for some clinical conditions [8]. Clinical studies support use of acupuncture as an integrative therapy for various medical conditions including postoperative nausea and vomiting, addiction to pain medication after dental surgery, stroke rehabilitation, headache, menstrual cramps, tennis elbow, fibromyalgia, myofascial pain, osteoarthritis, low back pain, carpal tunnel syndrome, and asthma as supported by the NIH consensus statement [9].

Although the efficacy, safety, and acceptance of acupuncture for pain management are well established among the adult population [10–21] (Table 23.1), evidence of its use for the treatment of pediatric conditions is limited [22]. The lack of pediatric data has been linked to the perception of fear of needles among children, which has created a barrier to acceptance of acupuncture in the pediatric population. There are a few studies demonstrating that in many cases children are open to this intervention, especially for chronic illnesses [23–25], and that this fear may be overcome by careful explanation and demonstration before introducing acupuncture therapy. This is further supported by the fact that more than 30 % of pediatric pain centers in the USA offer acupuncture therapy as a treatment modality [26, 27].

Table 23.1 Systematic reviews on acupuncture and pain disorders

Disorder	Review	No. of trials	Results
Low back pain	Manheimer et al. [10]	33	Positive
	Furlan [11]	35	Positive
Osteoarthritis	Manheimer et al. [12]	13	Positive/ inconclusive
	White et al. [13]	13	Positive
Dental pain	Ernst [14]	16	Positive
Headache	Melchart et al. [15]	26	Positive trend
Chronic pain	Patel et al. [15]	14	Inconclusive
	Ter Riet et al. [17]	51	Inconclusive
	Ezzo et al. [18]	51	Inconclusive
Fibromyalgia	Berman et al. [19]	7	Positive
Elbow pain	Gree et al. [20]	4	Inconclusive
	Trinh et al. [21]	6	Positive

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Functional Disorders

As described earlier in this book, children and adolescents presenting with physical symptoms and complaints that are medically unexplained after routine medical assessment are commonly categorized as having functional disorders. These diagnoses can be both conceptually and practically puzzling to the healthcare professionals. The symptoms reported by these patients are subjectively physical, yet objectively without explanatory pathology, and are often associated with anxiety and depressive symptoms and disorders [28]. Functional gastrointestinal disorders such as functional abdominal pain, functional dyspepsia, constipation, and irritable bowel syndrome are reported to have the highest incidence among the functional disorders (Chap. 5) and are some of the most common reasons for which children are treated with acupuncture. Functional abdominal pain usually has a significant negative impact on a child's life, resulting in more school absences per year than reported in healthy children. It may take an average of 25.9 months before a diagnosis is made [29] and impacts the entire family. Besides worrying about their child's health, parents have to endure the increased costs of healthcare use, lost wages for time taken off from work, as well as possible disruptions in family plans and activities [30].

Routine use of any pharmacological agent as a first-line therapy for the treatment of functional abdominal disorders has little support in the published literature. A strong consideration in management of functional disorders should be given to a treatment plan that includes non-pharmacological therapeutic interventions [31].

Acupuncture therapy can prove to be a useful adjunct intervention for management of functional disorders as shown by published literature in both the basic science and clinical studies [32, 33]. Exploratory studies to examine effects of electroacupuncture (EA) on gastrointestinal secretion, sensation, motility, and myoelectrical activity have been conducted in both humans and animals [34, 35]. EA has been shown to be associated with decreased basal acid output in healthy volunteers. It also reduced the vagally mediated acid output as demonstrated by reduction in acid production with sham feeding, while it had no effects on the pentagastrin-stimulated acid output [36]. EA was also effective in protecting the stomachs of rats with stress-induced gastric ulcer by thickening gastric mucosal barrier, stabilizing mast cells, and decreasing the gastrin level in gastric mucosa [37].

Efficacy of EA for the treatment of gastrointestinal motility disorders, and improvement in gastrointestinal symptoms, has been reported in patients with various disorders associated with gastrointestinal motility [38–41].

Enuresis and constipation in the absence of objective findings are deemed to be functional disorders and can affect the patient and their family's quality of life significantly, not to mention the social stigma that is associated especially with enuresis and encopresis. Acupuncture has been demonstrated as an effective intervention for management of these disorders [42–47].

Acupuncture may also be an effective intervention alone or as an adjunctive therapy for the treatment of insomnia. Results from a recent Cochrane systematic review reveal that, compared with no treatment or sham/placebo acupuncture, acupuncture resulted in more people with improvement in sleep quality [48].

Practical Application

Pediatricians and other primary care physicians are in an excellent position to identify patients who may benefit from acupuncture treatment. These patients are often children whose symptoms do not completely respond to conventional Western treatment regimens or whose therapeutic responses are limited as a result of side effects related to the therapy. Additionally, children of parents who themselves utilize acupuncture or have an interest in acupuncture therapy may be good candidates. Although the practice of acupuncture often requires specific training and certification, comparatively, *acupressure* requires minimal guidance and education and may be more readily practiced by a primary care provider, parents, other caregivers, or the children themselves.

Acupressure is a technique for stimulating the acupuncture points using digital pressure instead of needles or electrical or laser stimulation in order to achieve the therapeutic benefits. Like acupuncture, acupressure therapy is believed to be effective in the relief of pain, stress, and anxiety-related ailments and is ideal for self-treatment. It may also provide the patients or their caregivers with tools to implement symptom management in between the practitioner-delivered acupuncture treatments. The ideal degree of digital pressure needed for achieving treatment effects has not been defined but is believed to be equal to pressure needed to blanch the nail bed of the person applying acupressure, resulting in a feeling of soreness or achiness but not frank pain. Similarly, the ideal time period needed for effective acupressure also has not been defined and is variable depending on the age of the patient and indication for acupressure. For example, for pain and stress, firm continuous pressure is better, whereas for spasm-related discomfort, short bursts of firm pressure are recommended [49]. In general, pediatric patients need less time for acupuncture or acupressure. A brief period of acupuncture or acupressure, 5–10 s in neonates and children, may be effective for the

management of symptoms [48–50]. Broader guidelines recommend digital pressure at acupuncture points for periods of 20–30 s at a time for a total period of 2–5 min and can be repeated 2–3 times a day for more effective treatment of ongoing or chronic problems.

The efficacy of acupuncture has been shown to be dependent on the intensity of stimulation, i.e., the therapeutic response to EA stimulation is more intense compared to that of manual needle stimulation or acupressure. Often, acupuncture therapy may be necessary to obtain the optimal therapeutic response, but this response can be sustained with resultant prolonged therapeutic benefits, if patients or their caregivers are provided with directions for acupressure.

How to Identify a Practitioner

There are over 2,500 medical acupuncturists currently practicing in the USA. Medical acupuncturists are physicians with specific training in acupuncture in addition to their traditional medical training, and they often practice acupuncture in concert with their medical specialty, such as a pediatric anesthesiologists who complement their pain medicine practice with practice of acupuncture. Licensed acupuncturists (LAcs) undergo specific training for a period of 3–4 years and are required to meet licensing standards in the state in which they practice. Although there is no subspecialty training in pediatric acupuncture, some medical acupuncturists and LAcs incorporate the treatment of children in their acupuncture practice and thus may have more expertise in managing this challenging population. As with any referral, a discussion with the practitioners regarding the demographic makeup of their patient population and scope of their clinical practices may define their suitability for a referral source. Some insurance carriers cover acupuncture services; however, it is prudent for the patient or parent to query their individual policy, as coverage varies widely.

Case Studies

Case 1

Chronic Abdominal Pain

A 13-year-old girl presented to her pediatrician with increasing symptoms of abdominal pain. She noted that bouts of abdominal pain began about 3 years prior to presentation and now had progressed to become constant. She localized the pain to the periumbilical region and described it as continuous and sharp pain without burning sensation. She noted that she had difficulty with falling asleep and also reported intermittent symptoms of bed-wetting. Her past medical history was otherwise normal.

Under consultation with a urologist, she had undergone a series of diagnostic studies including renal ultrasound and urodynamic studies to evaluate her persistent enuresis. Additionally, a pediatric gastroenterology consultation was obtained, and she underwent an upper GI series, CT scan of the abdomen, colonoscopy, and cystoscopy to evaluate her abdominal pain. All results were unremarkable. Blood work included a CBC and serum electrolytes, which also were unremarkable.

Her medication regimen at presentation included polyethylene glycol (Miralax), fluoxetine (Zoloft), amitriptyline (Elavil), and oxybutynin (Ditropan). Despite this regimen, her symptoms persisted.

Physical examination revealed a pleasant, shy, right-handed girl in no acute distress. Vital signs were within normal limits. Her abdomen was soft, non-distended, and mildly tender upon palpation in all quadrants without evidence of rebound tenderness. Her bowel sounds were normal.

She was referred to the medical acupuncture service for treatment of her abdominal pain. Although the patient herself had never received acupuncture, her mother had reported excellent therapeutic results with the use of acupuncture, as she suffered with headaches. Weekly acupuncture treatments were conducted using needles at a combination of points (Fig. 23.5) including

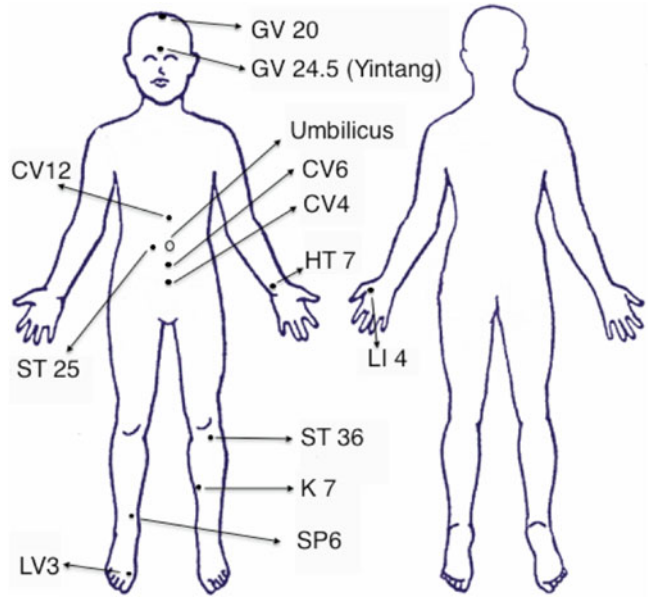

Fig. 23.5 Acupuncture points used for treatment in the case studies. *GV* Governing Vessel, *CV* Conception vessel, *ST* Stomach, *SP* Spleen, *LV* Liver, *KI* Kidney, *LI* Large intestine, *HT* Heart

Stomach 36 (ST 36), Spleen 6 (SP6), Conception Vessel 4 (CV 4), Conception Vessel 6 (CV 6), and Kidney 7 (KI 7). The duration of each treatment was 15 min. Over the course of several weeks, acupuncture treatment yielded short periods of relief followed by quick return of symptoms. During one of the acupuncture sessions, however, needling of two additional acupoints [Governing Vessel 20 (GV 20) and Heart 7 (HT 7)] was added to assist with calming and release of tension. With these added points and during the treatment, the patient expressed a significant cathartic emotional release of "worries" including a long-standing fear of losing her mother. Her mother revealed that several years earlier, she (the mother) had a prolonged course of fungal sepsis after lobectomy for treatment of lung cancer. While her mother was admitted to the ICU, this patient was cared for by her grandmother. The patient expressed a daily fear that her mother had passed away when she returned home from school.

Acupuncture is used commonly as a physically oriented therapy, but occasionally, it can aid with highlighting mental health diagnoses as it may uncover underlying psychological or emotional symptoms in the course of its application. When acupuncture was rendered as a treatment for physical and emotional relief in this patient, it allowed the patient to express her repressed feelings and fears that seemed to have been contributing to her physical symptoms.

Questions

1. The most common cause of abdominal pain in children is:
   (a) Intestinal obstruction
   (b) Gastritis
   (c) Duodenal ulcer
   (d) Unknown etiology
   (e) None of the above
2. When is the optimal time to refer a child to an acupuncture clinic?
   (a) Immediately upon presentation.
   (b) Never refer a pediatric patient for acupuncture.
   (c) When a comprehensive evaluation has been done to investigate etiology.
   (d) When standard medical treatment is not helpful.
   (e) a and d.
3. Should you continue to treat her with acupuncture after her "cathartic experience?"
   (a) Yes, acupuncture may be used as an adjunct to psychological therapy.
   (b) No, the patient should be admitted to an inpatient unit for intensive evaluation and treatment.

(c) Never use psychiatric treatment and acupuncture together.
(d) Use acupuncture only to treat symptoms.

Answers

1. (d): Although intestinal obstruction, gastritis, and duodenal ulcer can cause abdominal pain in children, in most cases, the exact etiology remains unknown despite extensive workup.
2. (c): Referral to an acupuncturist can be considered at any point including when the patient initially presented for symptom management, but the optimal approach is to first establish the etiology of abdominal pain. Thus, acupuncture therapy should be considered once a comprehensive evaluation has been carried out so that any pathology responsive to medical therapy can be addressed along with symptom management.
3. (a): Acupuncture in this case was used to both relieve pain and promote emotional release thus aiding in uncovering psychological distress as the underlying etiology of the patient's pain. Acupuncture therapy in this case would be a good adjunct for both management of her physical symptoms (abdominal pain) and her psychological symptoms thus providing a comprehensive approach to her treatment.

Case 2

Headache

A 15-year-old girl presented to the medical acupuncture service with a chief complaint of persistent headache several months after a whiplash-like injury sustained while riding on a roller coaster. Initially, she was treated by her pediatrician, but as her symptoms persisted, she was referred to a neurologist. She underwent an extensive workup including a neurologic examination, magnetic resonance imaging, magnetic resonance angiography, lumbar puncture, and blood work to rule out infectious and autoimmune etiologies. Her neurologist diagnosed her as having a combination of chronic daily headaches and migraine with aura. She described her headache as an "allover" pain with spikes akin to a "thunderclap." She reported worsening of her headache with exposure to bright lights but no sonophobia. She was no longer attending school because of the pain.

At presentation to our clinic, she was being treated with verapamil, after failing to respond to a multitude of other prophylactic and abortive analgesic medications including acetaminophen, ibuprofen, sumatriptan, amitriptyline, and topiramate. Additionally she had tried several other interventions including botulinum toxin injections and an occipital nerve block, which she stated exacerbated her headache. She also underwent "cervical manipulation," which was mildly helpful, but this was discontinued with her recent referral. She reported that she had undergone one acupuncture treatment, which gave her some relief, and wanted to pursue this avenue for further pain relief. Her past medical and surgical history included a musculoskeletal anomaly of bilateral diminutive thumbs for which she had previously undergone a right first finger to thumb transposition for increased function.

On physical examination, she was a pleasant young woman in mild distress and preferred a darkened examination room as a result of her photophobia. Cranial nerve examination was unremarkable except for an absence of sense of smell (CN I). Although she complained of significant pain in her head to the point that she did not want to move or touch her hair, no allodynia was noted on examination. There was moderate tightness over the trapezii bilaterally. She had bilaterally absent radial pulses, a well-healed scar from her right-hand surgery, and a left thumb which had remained small.

She was treated with an acupuncture regimen with manual needle stimulation [Governing Vessel 20 (GV 20), Yintang or Governing Vessel 24.5 (GV 24.5), and bilateral Large Intestine (LI 4) (Fig. 23.5)], as well as cupping over the course of several weeks, and she tolerated the treatments well. She reported a significant decrease in her baseline headache (from an initial pain score of 12/10 to a 4/10) lasting for several days to a week after each treatment.

Questions

1. When is a headache classified as chronic daily headache?
   (a) When the headache is present every day
   (b) When the headache has no known etiology
   (c) When the headache occurs more than 15 days of the month for at least 3 months with no identifiable etiology
   (d) When there is no aura
2. When is acupuncture a more appropriate therapy for headache than abortive analgesics?
   (a) When the headache is very frequent
   (b) When the headache is severe in intensity and occurs suddenly
   (c) Only when a patient has a migraine headache
   (d) None of the above
3. Could acupressure have been as effective as acupuncture for this patient?
   (a) Yes, definitely
   (b) Definitely not
   (c) Perhaps

Answers

1. (c): Chronic daily headaches are defined as headaches that occur more than 15 days of the month for at least 3 months either with no identifiable etiology (primary) or due to another etiology (secondary) such as medication overuse, trauma, transformed migraines, etc. [51].
2. (a): Most headache experts recommend that the use of abortive analgesics should be limited for those headaches that are infrequent and their use should be limited to no more than 2–3 times per week. Any headaches that are more frequent in occurrence or that are more persistent such as migraines and chronic persistent daily headaches may require prophylactic therapy, including acupuncture.
3. (c): Acupuncture and acupressure work by the same basic principles; however, with acupressure, no needles are used to puncture the skin. Acupressure may be ideal for patients with contraindications to needle therapy (infection at the site of needling, bleeding diathesis, extreme fear of needles, etc.). This patient, as many children do, tolerated the needles well. In this setting, it makes sense to provide her with an acupuncture treatment using manual needle stimulation as it is believed that the strength of stimulation results in a better therapeutic impact (i.e., electroacupuncture > manual needle acupuncture > acupressure). However, had she been not able to tolerate the needles, acupressure may have been an appropriate option.

Case 3

Constipation

A 2-year and 4-month-old male was referred to our acupuncture clinic by his primary care provider upon the parents' request for treatment of chronic constipation. He was an otherwise healthy male who was born at term via cesarean section due to maternal history of preeclampsia. He did not require any supplemental oxygenation or special care in the postnatal period. His developmental milestones were reported normal to slightly early with walking at 9 months. He was very articulate for his age. He was fully potty trained by the age of 18 months. His parents reported that he had constipation for the past 6 months wherein his bowel movements had been occurring every 4–5 days despite use of laxatives and were associated with hard stools that occasionally were blood streaked and associated with crying during defecation. He did not exhibit any evidence of abdominal pain except for just before and during bowel movements. He remained playful and socially engaged. No sleep disturbance was reported.

He had been evaluated by his primary care physician, a gastroenterologist, and a surgeon to identify an etiology for his constipation. His workup had included a normal stool exam, abdominal X-rays, and CT scan that showed stool mass. Surgical assessment had also ruled out possibilities of Hirschsprung's disease, anal fistulas, or fissures. He had been tried on high-fiber diet, increased water intake, and elimination of juices and lactose from his diet without any improvement in his constipation. He had also been treated

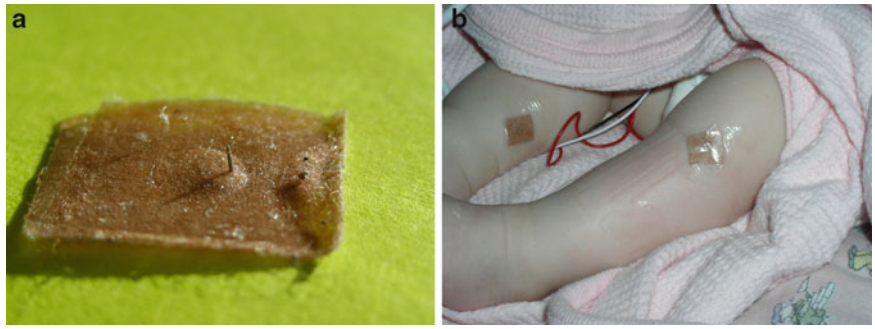

Fig. 23.6 (**a**) A press needle. (**b**) An infant receiving acupuncture treatment with press needles

with lactulose, polyethylene glycol (Miralax), and suppositories; none of which increased his bowel movement frequency.

Review of growth and developmental history revealed a brief period of colic when he was 3–5 months old and a brief history of constipation associated with painful defecation at the age of 1 year that resolved easily after a brief course of laxatives.

Physical examination revealed a cheerful young boy who was quite articulate and engaged easily with the provider. Vitals signs were normal. Abdominal examination revealed a soft, non-tender, non-distended abdomen with some palpable stool mass in the left lower quadrant. A physical examination was done specifically to make an acupuncture-based diagnosis and determine the acupuncture points to be treated (acupuncture examination). This revealed a pattern of the radial pulse characterized as "bounding" especially in the liver meridian position. (There are 3 positions on the wrist on each side corresponding to various meridians.) Also, there was a warmer middle part of the abdomen or middle jiao. (This is based on the Traditional Chinese Medicine diagnosis theory where the abdomen is designated as a "burner" and source of flow of heat and warmth indicating flow of qi. The abdomen is divided horizontally into three equal sections: upper jiao (section), middle jiao, and lower jiao.)

Weekly acupuncture treatment was conducted using press needles which are 1–2 mm long needles attached to an adhesive band-aid (Fig. 23.6a, b). Press needles were inserted at a combination of points including Stomach 36 (ST 36), Spleen 6 (SP6), Liver 3 (LV3), Large Intestine (LI4), cupping at Stomach 25 (ST 25), and Conception Vessel 12 (CV12) (Fig. 23.5). Duration of each treatment was 10–15 min. During some of the treatments, cupping was followed by application of press needles at ST 25 and CV12 with directions to the parents to remove them at home later that evening. After the third treatment, the parents reported that their child was having bowel movements every other day without the use of a suppository. At this point, the parents were also instructed in acupressure at ST 36, LI4, and LV3. After 6 treatments, they had reduced his Miralax dose to half, and he continued to have regular bowel movements every other day. The stools were softer in consistency. The acupuncture treatments were then spaced out to every other week for 2 months and eventually stopped (as was the Miralax), but parents reported using acupressure with continued resolution of his constipation.

Questions

1. Constipation in children:
   (a) Can be defined as lack of daily bowel movements
   (b) Can be defined as absence of liquid stools
   (c) Should be immediately referred to a gastroenterologist for further workup
   (d) Most commonly is due to willful avoidance of toilet
   (e) Is common among formula-fed children

2. Use of acupuncture in children:
   (a) Is contraindicated under the age of 6 months
   (b) Requires special training in pediatric acupuncture
   (c) May require careful explanation, demonstration of acupuncture, and distraction techniques
   (d) Helpful for treatment of painful conditions only
   (e) Should be the first line of therapy
3. The duration of acupuncture therapy is determined by all *except*:
   (a) Etiology of symptoms
   (b) Duration of symptoms
   (c) Patient's desire
   (d) All of the above
   (e) None of the above

Answers

1. (d): Constipation is defined as having a bowel movement fewer than three times per week. A frequency of bowel movements occurring three times a day to three times a week constitutes normal bowel habit. However, with constipation, stools are usually hard, dry, small in size, and difficult to eliminate. Children often develop constipation as a result of willful stool withholding because they are stressed about potty training, are embarrassed to use a public bathroom, do not want to interrupt playtime, or are fearful of having a painful or unpleasant bowel movement. Constipation is common in children and is usually without long-term consequences; however, it can diminish a child's quality of life, cause emotional problems, and create family stress. Rarely, constipation is a sign of a more sinister health problem. Withholding of stool may result in hard, dry, and difficult to pass feces and sometimes even fecal impaction. Stool builds up behind the impaction and may unexpectedly leak, soiling a child's underwear, and may be often mistaken by parents as a sign of diarrhea. When a child's abdominal pain is also associated with fever, emesis, blood in stool, abdominal distension, significant weight loss, anal fissures, etc., it heralds presence of more ominous etiology and would warrant further investigation and a referral to a gastroenterologist or even an emergency room in case of acute abdomen.
2. (c): It is not uncommon for children to experience some degree of needle phobia which can make them apprehensive about acupuncture treatment. However, the literature supports that use of careful explanation, demonstration of acupuncture, and distraction techniques are very successful in pediatric patients. Occasionally, demonstration of acupuncture on the parents, the children's favorite toys, or the patients themselves helps overcome their apprehensions. Although experience working with children is important and extremely useful for acupuncturists who treat children, there is no special training requirement set forth for practice of pediatric acupuncture.
3. (d): The duration of acupuncture therapy is dependent on several factors including the etiology of a disease, duration of symptoms or chronicity, patient's response to treatment, as well as a patient's acceptance of the treatment and ability to follow up with an acupuncturist. Usually acute illnesses or events will require a brief period of treatment such as in case of postoperative nausea or vomiting, while chronic conditions will require acupuncture therapy for several weeks to months such as in case of chronic headache or back pain. Seasonal symptoms such as allergic rhinitis may require a series of initial treatments followed by intermittent maintenance therapy corresponding to the surge of allergens in the environment. Usually, for a chronic condition, a series of 8–10 treatments 1–2 times weekly is recommended to evaluate the efficacy of acupuncture therapy for that condition. If a patient does not benefit from acupuncture therapy in this time period, it is unlikely that continuing acupuncture beyond that point would have any additional benefit. If the patient has a therapeutic response in this time period, the frequency of treatments is then decreased gradually until the patient is able to manage the symptoms without regular acupuncture treatments. Maintenance therapy may

be required for some ailments as mentioned earlier in case of allergic rhinitis. A patient's ability to tolerate acupuncture therapy or make regular acupuncture therapy sessions either due to scheduling issues or financial issues may impact the duration of treatment.

Conclusions

Acupuncture and acupressure are established and effective treatment modalities that can be easily incorporated into an integrative medical management regimen, with minimal adverse outcomes. Evidence for their roles as therapies to complement the treatment regimen for functional symptoms is emerging. However, their roles need to be investigated through more systematic and controlled studies. It is helpful for physicians to understand the potential role that acupuncture and acupressure may play in the management of their patients.

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Mindfulness Meditation for Children

24

Carisa K. Perry-Parrish and Erica M.S. Sibinga

Abstract

Mindfulness meditation has been described as a useful adjunct to behavioral and medical interventions in the effective management of a range of somatic symptoms, including abdominal pain and atopic dermatitis. Mindfulness approaches can be taught to children, adolescents, and their parents to help reduce stress processes that result from and/or exacerbate functional symptoms. Moreover, mindfulness parenting techniques can augment traditional behavioral approaches to improve children's behavior through specific forms of mindful parent–child interactions. A growing body of empirical studies and clinical experience suggest that the incorporation of mindfulness meditation will enable clinicians to more effectively treat pediatric patients and their families in coping optimally with a range of challenging physical symptoms. This chapter provides a description of mindfulness approaches, theoretical mechanisms of action, literature review, a sample script for a beginning mindfulness exercise, and case studies.

Keywords

Abdominal pain • Atopic dermatitis • Mindfulness • Meditation • Parent–child special time

Background

Mindfulness has been described as "paying attention in a particular way: on purpose, in the present moment, and nonjudgmentally" [1]. In essence, mindfulness is the complete awareness of what is happening right now or "being in the zone." Therefore, mindfulness instruction is intended to enhance an individual's innate ability to be aware. Formal mindfulness instruction entails a range of techniques that help foster an intentional focusing of attention on one's

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present-moment experience while letting go of negative, self-critical judgments. As detailed explicitly in many mindfulness programs, this type of training aims to help individuals accept unpleasant and painful experiences without reactively attempting to change the experience [2]. However, as most of us would prefer to reduce or eliminate pain and discomfort as much as possible, some mindfulness-based programs additionally adopt a dialectical position of balancing desire for change alongside intentional acceptance of the inevitability of suffering [2]. As moment-to-moment awareness through the day is the ultimate goal of mindfulness programs, there is also instruction of informal techniques that can be used at any time.

The application of mindfulness meditation to reduce suffering has been a feature of behavioral medicine in the West for well over 30 years [3]. Meditation practices complement a group of established psychological approaches to reduce stress and discomfort, including cognitive-behavioral therapy (Chap. 19) and relaxation techniques. Mindfulness-based stress reduction (MBSR) is one of the more well-known programs, which initially was developed for use with adults presenting with chronic pain and other chronic and stressful conditions [3]. Since the establishment of MBSR, several mindfulness-based interventions have been developed and applied to treat a range of psychological problems among adults [4], including dialectical behavior therapy (DBT) [5], acceptance and commitment therapy (ACT) [6], mindfulness-based cognitive therapy (MBCT) [7], and mindfulness-based relapse prevention (MBRP) [8]. The common thread among these other approaches is a dual emphasis on mindfulness and behavioral change as core processes to alleviate suffering.

An information-processing model has been proposed [9] that identifies several ways in which mindfulness could work by modifying attentional and emotional self-regulation processes: (1) interrupting automatic, "mindless" habits and cognitive scripts associated with maladaptive behavior; (2) changing an individual's relationship to his or her own memory activation (e.g., neutrally observing a memory, rather than attempting to inhibit it or reacting emotionally in a negative way); (3) becoming desensitized to previous emotional triggers for behavior; and (4) developing increased attention to and awareness of one's own cognitive and emotional processes generally. It has been theorized that mindfulness "may change automatic response tendencies when the patient observes, describes, and participates in emotional experiences without acting on them" [10]. Indeed, the proposed mechanism of therapeutic change in DBT is the reduction of ineffective action tendencies that are linked with emotion dysregulation [10]. Similarly, reduction of psychological inflexibility and experiential avoidance within ACT theory are proposed to allow individuals to observe their psychological experiences instead of attempting to control them [11]. This psychological shift may promote a number of associated cognitive, emotional, and behavioral changes (see Table 24.1, for examples).

Table 24.1 Potential psychological changes associated with mindfulness training

Reduced belief in automatic thoughts
Increased flexibility of attention
Improved intentional shifts in attention
Reduced emotional intensity/duration and secondary emotional responses
Enhanced ability for learning
Enhanced treatment motivation

Mindfulness training is theorized to result in improved self-regulation that emerges from increased acceptance and self-awareness, such as noticing unpleasant emotions and distress as experiences that can be accepted, rather than impulsively reacted to, ruminated over, or chronically avoided in an ineffective manner [4, 12, 13]. This enhanced acceptance of one's internal experiences is thought to lead to reduced suffering and distress in response to stress. Thus, mindfulness training may result in symptom reduction through exposure to emotional and psychological sensations, changes in attitude/cognitive stance, greater use of self-regulation and coping skills, and acceptance of psychological experiences [4].

Additionally, behavioral and learning theory may also explain the potential utility of mindfulness-based interventions for treating somatic symptoms. First, mindfulness-based

interventions may reduce emotion regulation problems that precipitate and follow social/interpersonal conflicts and stressors that often function as triggers for exacerbations in somatic symptoms. Second, mindfulness may also reduce high arousal and emotional reactivity from which individuals may seek immediate relief, thus counteracting the negative reinforcement value that functional somatic symptoms may have previously provided. Thus, in many ways, mindfulness may address maladaptive homeostasis, self-soothing, and avoidant responses as evidenced by studies among adults with somatic difficulties.

Despite decades of research in adults, there are relatively few published studies of mindfulness-based interventions for use with children. However, this small literature provides preliminary evidence that mindfulness-based treatments are feasible and beneficial for use in pediatric populations (for reviews, see [14, 15]). A number of mindfulness programs have been adapted for use with children and youth. Adaptations typically involve shortening the formal mindfulness techniques when they are introduced, with a gradual increase in duration as the course progresses; clarifying and concretizing language used for instruction; and providing age-appropriate mindfulness activities. A randomized trial of MBSR compared with usual care for adolescents in outpatient psychiatric treatment showed significant reductions in anxiety and depression and improvements in global psychiatric functioning [16]. In a study of substance-abusing adolescents [17], MBSR was well tolerated and accepted and seemed to effectively complement other therapeutic components (sleep hygiene, stimulus control, and cognitive therapy) in reducing sleep problems.

DBT is a cognitive-behavioral therapy that incorporates mindfulness to treat individuals with emotional dysregulation. DBT involves four major treatment modules: mindfulness, distress tolerance (e.g., self-soothing, radical acceptance), emotion regulation, and interpersonal effectiveness (e.g., communication, social goals). DBT has been modified for use with adolescents (DBT-A) to include an emphasis on improving parent–child interactions. Among adolescents with oppositional defiant disorder, 16 weeks of DBT-A skills training was shown to reduce self- and parent-reported internalizing and externalizing symptoms [18]. A year-long trial of DBT-A involving family skills training and individual therapy in a small sample of adolescents with bipolar disorder demonstrated feasibility and acceptability of the treatment as well as indicated significant improvements in feelings of suicidality, self-harm, emotional dysregulation, and depressive symptoms [19]. DBT-A was well received among adolescents with severe emotional dysregulation (e.g., significant self-harming behavior), with mindfulness and distress tolerance skills being rated as particularly helpful components of the intervention by adolescents [20]. DBT-A has been shown to reduce behavioral problems among incarcerated adolescent females with mental health problems, with a majority also presenting with comorbid substance abuse [21]. In addition, implementation of DBT-A resulted in decreased premature terminations from residential treatment facilities, due to self-harm and psychiatric hospitalization, and reduced number of days spent in psychiatric hospitals among adolescent young women [22].

A small feasibility trial of mindfulness-based cognitive therapy for children (MBCT-C) found support for acceptability and reduction of internalizing (e.g., anxiety, depression) and externalizing symptoms (e.g., disruptive behavior) among a non-referred sample of preadolescents [23]. Studies of MBSR for youth recruited from an urban outpatient primary care clinic have shown program acceptability, feasibility, and benefit related to improved relationships and coping, and reductions in conflict engagement, anxiety, and stress [24, 25, 26]. In high school students, mindfulness instruction led to improvements in elevated blood pressure [29]. Additionally, mindfulness instruction (with age-appropriate adaptations, such as belly breathing, focusing on breath, and the use of "mind jars") has been studied in younger students, showing benefits in attention and executive function [27, 28].

In summary, although the literature is limited, evidence suggests that mindfulness-based interventions are beneficial for children and adolescents

to enhance their self-regulation and coping, which are aspects central to the management of functional symptoms.

Practical Application: A Simple Mindfulness Meditation Practice

Many clinicians, who teach mindfulness techniques across a variety of empirically supported approaches such as MBSR, ACT, DBT, and MBCT, emphasize the need for the practitioner to gain personal experience with mindfulness meditation. Experience in mindfulness then enables the practitioner to help children and adolescents learn through their own experiences what mindfulness is and is not and how mindfulness techniques can be used to cope with a range of stressors. Providing a definition of what mindfulness is can be a helpful first step. In our work, we often use Jon Kabat–Zinn's classic conceptualization, in which mindfulness is simply directing attention in a particular way: on purpose, in the present moment, and nonjudgmentally. By contrast, mindfulness is the opposite of mindlessness or being on "autopilot" [1].

Mindfulness training on one's breathing is a traditional starting point in meditation. The following text is a sample script for a brief meditation on one's breath:

1. *Sit in a comfortable position in your chair. Sit upright, with your back straight* (but not uncomfortably so) *and your feet flat on the floor*.
2. *Gently bring your attention to your breath. There's no need to change how you are breathing in any way, but just notice each breath as you inhale and exhale.*
3. *Be aware of the sensations in your body as you breathe; notice the sensation of the in-breath as it enters your nose, throat, lungs; notice your out-breath as you exhale.*
4. *Observe what parts of your body move as you breathe. Your shoulders, chest, stomach may move. Perhaps other parts of your body move too—such as your arms and feet.*
5. *If other thoughts come in to your mind as you practice, acknowledge them and then gently shift your awareness back to your breathing again. It is okay to notice these other thoughts; let them go and return your attention to your in-breath and out-breath.*
6. *Continue for 2–3 min or more, as you like.*
7. AFTER: *What did you notice? What was it like?*

Although this is a simple mindfulness exercise, it may not be easy to maintain focus on the breath; this is common and reassurance that lots of practice is needed can be given. Also, it allows what may be a very different experience than a typical moment spent worrying about whether an uncomfortable feeling will not end or ruminating about how one is frequently bothered by an unwanted experience (e.g., itching in atopic dermatitis, abdominal upset/pain). This simple exercise can be modified and expanded in a number of ways. For example, as one breathes in, a simple phrase to describe that action could be added ("*I am breathing in my breath, I am breathing out my breath")*. Likewise, a general coping statement could be added (e.g., *I breathe in peace, I breathe out stress*) or an observation of undesired sensations could be noticed and described nonjudgmentally (e.g., *I notice an itch on my arm*). As Thich Nhat Hanh writes in *The Miracle of Mindfulness,* "Mindfulness frees us from forgetfulness…and enables us to live" [30]. Since enhancing awareness of what is happening in the present moment is the intention of mindfulness instruction, discussion of what *actually* happens during such an exercise provides an opportunity for the patient to gain understanding of how his or her mind "works." Practice of such techniques allows for more facility in that awareness and greater ability to manage one's attention.

Repeated over days and weeks, the goal of mindfulness training is to help individuals stay in their present experience, whether enjoyable or uncomfortable, and see things nonjudgmentally and clearly as they are. Seeing one's experience clearly means not catastrophizing (i.e., seeing things worse than they are) and not denying (i.e., ignoring when things are truly undesirable). Rather, purposefully staying in the moment, with full, clear attention and without judgment, can lead to many positive outcomes for psychological and physical health. Mindfulness instruction provides simple techniques to "check in" with oneself

to assess what is actually happening in the present moment. This "checking in" often leads to enhanced perspective taking [25, 26, 31] and enhanced self-regulation [32]. While mindfulness techniques can enhance individuals' abilities to see their present-moment lives more clearly, it is typically believed to be a lifelong practice.

Case Studies

Although MBSR has been studied in a number of adult medical conditions (e.g., chronic pain, psoriasis, inflammatory bowel syndrome), there are fewer such studies among children. Application of mindfulness training targets the role of stress in the mind–body connection. Many conditions implicate a role of stress in exacerbating or maintaining dysfunctional somatic problems.

Atopic dermatitis—Childhood atopic dermatitis affects up to 20% of youth under the age of 5 years. Pruritus, or severe itching, is a core symptom of atopic dermatitis flares. Despite the availability of effective treatments, many children and parents can become quite distressed in trying to manage and reduce the distressing intensity of the itch sensation that accompanies periods of eczema flare (Chap. 7).

Case #1

Max is a 12-year-old male with a long-standing history of atopic dermatitis. He has suffered for many years with extreme itchiness, which has disrupted his sleep, interactions with parents, and academic and social functioning at school. In addition to frequent emollient use, he has been prescribed a range of topical medications to address his skin inflammation, which do seem to be effective. Unfortunately, he seems to have developed habitual scratching in response to any pruritus.

Questions

1. On exam, his skin appears mildly dry diffusely, but no areas of erythema or excoriation suggestive of a flare. It seems that his scratching behavior is quickly triggered by any itching sensation, no matter how mild. Based on history and exam, it appears his atopic dermatitis is well controlled on the current regimen. What is the most reasonable next step?
   (a) Add oral steroids to escalate atopic dermatitis treatment.
   (b) Start him on a daily antibiotic to treat presumed superinfection.
   (c) Discontinue his topical treatments entirely.
   (d) Refer him for behavioral management of his response to pruritus.
   (e) Refer him for psychiatric treatment.
2. What are the goals of the above choice?
   (a) To have him evaluated for hypochondriasis.
   (b) To eliminate the skin bacteria that is causing the persistent pruritus.
   (c) Oral steroids are often beneficial for atopic dermatitis.
   (d) To simplify his treatment regimen, so he can forget he has a skin condition.
   (e) To improve his ability to cope with the sensation of pruritus.

Answers

1. (d): Behavioral techniques, such as habit reversal training and positive reinforcement of differential responses, can help young people overcome the overlearned habit of scratching in response to pruritus. Parents can be taught to coach children through these techniques to replace other, less effective parenting strategies to address scratching (e.g., nagging, punishing).
2. (e): Itching is a powerful stimulus that elicits scratching in most individuals. Unfortunately for some children with eczema, this signal elicits too much scratching that results in a pernicious itch–scratch–rash cycle that can overwhelm very effective medication treatments. In addition, some children develop a habit of scratching that is out of proportion to their condition or even in the absence of pruritus. Parents of these children comment that they appear to scratch when upset, frustrated, or stressed in some way.

In combination with behavioral strategies to reduce Max's scratching habit, mindfulness techniques helped him observe his experiences and his own habitual behavioral responses to them.

Max was taught with habit reversal techniques in which he engaged in a new behavior to cope with itching (e.g., clasping hands together) that was incompatible with scratching. Although Max had been practicing habit reversal, he had difficulty in engaging this strategy to prevent scratching. One mindfulness exercise was to observe the itch as it emerged and then receded over the course of 20 min. He was able to notice how the itch intensified and elicited a strong urge to respond by scratching. He also observed and described through this experience that scratching did not make the itch go away; rather, the itch resolved on its own. Although this was not described to Max as mindfulness, he understood that we were observing his experiences without judging (e.g., "this itch will never go away" or "I am bad for feeling this itch") or automatically reacting (e.g., through habitual scratching). Rather, Max was coached to simply notice and describe the experience (e.g., "the itch is getting itchier" or "the itch is less intense"). Through Max's direct-guided mindful observation of his present-focused experience, he was able to appreciate the natural course of an itch without reacting, which gave him confidence to resist similar urges to scratch. This instruction does not mean that Max will never itch, but it provided him with information and awareness about the nature of both the experience itself and his reaction to the experience, which will inform his future experiences.

Abdominal pain and nausea represent another common difficulty. Abdominal pain is frequently treated initially with medication, but families often present with children who continue to report pain and discomfort that hinders optimal functioning (Chap. 5).

Case #2

Jane is a 15-year-old female with a several-month history of diffuse abdominal pain and nausea. Her medical workup was negative. The symptoms seem to have emerged during a time of significant school stress. She was evaluated by a pediatric gastroenterologist, who treated her with acid suppression, but her pain persisted. Further workup by the gastroenterologist was "negative," and the family was assured that her symptoms did not represent physical pathology. Nevertheless, Jane had been missing a significant amount of school due to her abdominal pain and expressed concern about vomiting at school. Her pain and nausea disrupted her appetite, and her sleep schedule was thrown off since she sometimes stayed up late. Her symptoms and school absences were creating stress for her parents who felt at a loss to ease her discomfort and whose schedules were disrupted.

Questions

1. On exam, Jane is a well-appearing, pleasant girl, with an entirely normal physical exam. She says that she does want to return to school. What should you suggest?
   (a) The family should get a tutor for homeschooling.
   (b) She should work with a behavioral therapist or psychologist to learn how to cope with her symptoms.
   (c) Stick with the acid suppression.
   (d) Diagnose her with sleep disorder and prescribe her sleeping pills.
   (e) Refer her to a psychiatrist for diagnosis of social anxiety disorder.
2. What are the goals of the above choice?
   (a) Complete her high school education.
   (b) Medical treatment for social anxiety disorder.
   (c) Improve her ability to cope effectively with her symptoms.
   (d) Sleep disruption is the root of her symptoms, so they will resolve with sleeping medication.
   (e) Acid suppression will eventually be effective.

Answers

1. (b): Behavioral pain management approaches focus on identifying thoughts, emotions, and behaviors that exacerbate physical experiences and discomfort. Such cognitive-behavioral approaches may help individuals identify maladaptive coping patterns that can be altered

through stress reduction techniques, such as mindfulness.

2. (c): As it seems that Jane's symptoms are significantly interfering with her general well-being but do not represent a pathology requiring additional treatment, the goal is for Jane to learn how to cope better with her symptoms, so she can reengage with the activities of her life.

Jane's mindfulness training focused on observing her thoughts and working to change her relationship to those thoughts. In particular, she practiced noticing her thoughts that predicted the worst possible outcome (i.e., throwing up) and consciously attending to her actual experience in those moments that demonstrated the opposite (i.e., not throwing up). Over time, she was able to accept her thoughts as just that—thoughts—that did not necessarily reflect reality. As she became more experienced, her mindful stance toward her thoughts allowed her to see them as thoughts rather than facts. With these exercises, Jane was able to reduce her pain complaints and return to school without subsequent absences.

Special time—For children with somatic complaints and behavioral problems and even for healthy children, pediatric clinicians have long recognized the need to encourage parents to nurture positive relationships with their children through positive parental attention. The prescription for regular, recurrent "special time" has been a popular approach to this need. The practice of mindfulness in its fullest expression means living and attending to the present moment at all times, with implications for how we manage our lives and our relationships [33]. Although not typically thought of as a formal mindfulness technique, "special time" does function to increase awareness of the present-moment experience of the parent–child dyad and can be seen as supporting mindfulness in the parent–child relationship, particularly for the parent. "Special time" is a time when child and parent spend time together that is free from other tasks or interruptions such as televisions and cell phones [34]; a time that is fundamentally about being together with a present-focused attitude. Often, it is recommended that the child chooses the activity for the time together, that the parent brings an open and positive attitude, and that the parent be in-tune with their child for that period of time, however brief. Thus, the parent is instructed to be aware, open, positive, and attentive to the experience of being with her or his child as it unfolds in the present; the parent is instructed to be mindful.

Case #3

Jasmine is a 4-year-old whose parents complain of behavioral problems. Reportedly, over the past 2 months, she has been having tantrums and has become uncooperative with her previously well-established weeknight bedtime routine at 7:30 p.m.. When Jasmine goes to bed later, she has a very difficult time getting up in time for preschool in the morning, and her teachers say her behavior and mood are bad. Her father recently started a new job, with which he is delighted, but he does not get home until about 9 p.m.

Questions

1. Jasmine's physical exam shows an active and engaged 4-year-old with normal examination, normal growth, and development. What do you suggest?
   (a) Change Jasmine's bedtime to 10 p.m., so she can see her father when he gets home.
   (b) Coach her mother on "tough love."
   (c) Start her on diphenhydramine at 7:00 p.m. to help her sleep.
   (d) Prescribe "special time" with her father for Jasmine.
   (e) Rearrange her bedroom furniture.
2. What are the goals of the above suggestion?
   (a) Recognition that her behavior is keeping her up later anyway.
   (b) Respond to Jasmine's desire to have more time with her father in a way that's acceptable to all family members.
   (c) Reinforcing the mother's control of the situation.
   (d) Change up the routine to help her understand that things change sometimes.
   (e) Make Jasmine drowsy at the appropriate bedtime.

Answers

1. (d): Special time is a core component of most empirically based parent management approaches. Parents are coached to attend to their child and reinforce appropriate behavior through attention, observation, reflection, and praise. Put another way, parents are asked to engage in mindfully attending to their child and their child's behavior during daily activities and play. Further, special time typically results in improved parental appreciation of the child and improved parent–child relationship, as well as improved behavior [35]. Both children and parents benefit from the present-focused time together, particularly related to mutual appreciation and positive parent–child relationship.
2. (b): Due to competing demands for their time, many parents report feeling rushed and are frequently unable to focus on their children with undivided attention. In turn, children often display behavioral worsening that is inadvertently reinforced through negative parental attention. Thus, the goal of special time is to promote a more positive, loving, attuned parent–child relationship. Having a positive parent–child relationship is a crucial foundation for the success of subsequent behavioral approaches to changing misbehavior.

"Special time" was prescribed. Jasmine and her father started having breakfast at the table together daily before she went to preschool during the week and a 30-min time together on Saturday mornings. In this example, the parent was taught a mindfulness strategy to use within the parenting context to address behavioral issues and to nurture a positive parent–child relationship. Within a short period of time, Jasmine's tantrums decreased and 7:30 p.m. bedtime went smoothly again. While Jasmine's father was able to set aside specific times, many families find significant meaning in a more flexible approach to special time [36], perhaps drawing even more on principles of in-the-moment mindfulness.

In the cases reviewed here, mindfulness techniques were added to an array of medical and/or behavioral strategies for supporting improvement in functioning. While the concept of mindfulness may seem simple, its practice and the ability to provide quality mindfulness instruction are far from easy. As reviewed earlier, there are several psychological treatments that emphasize mindfulness training, including MBSR, MBCT, DBT, and ACT. Listed in the appendix of this book are websites that provide information about recommended readings and opportunity for training and certification in some of the specific mindfulness treatment modalities

Conclusions

Mindfulness meditation instruction has been shown to improve mental health and quality of life outcomes. Mindfulness instruction leads to reduced stress and enhanced self-regulation, which can be thought of as the intertwined processes of psychological functioning, cognitive functioning, and coping (Fig. 24.1). In particular, mindfulness has been found to reduce psychological symptoms, such as anxiety; improve

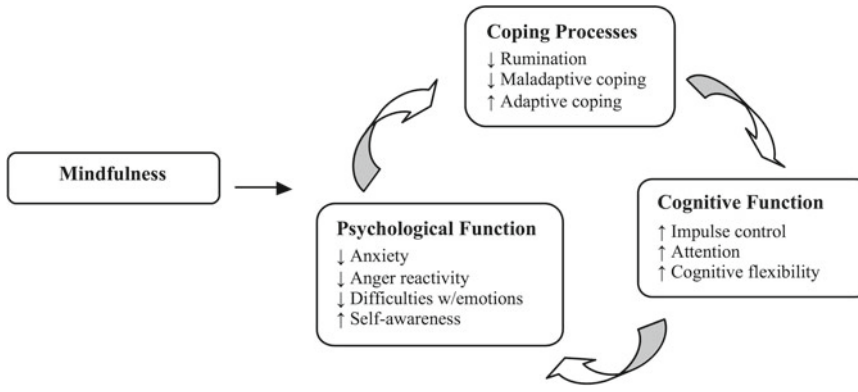

Fig. 24.1 Mindfulness and improved self-regulation

emotion regulation; improve attention and the ability to focus; and reduce maladaptive coping and rumination. These outcomes have been associated with increased calm, improved relationships, and reduced stress and anxiety.

Mindfulness techniques are likely to be especially beneficial for children suffering with functional symptoms, as it supports a positive change in their relationship to their symptoms. Enhancing the mindful awareness of the present-moment experience of the bodily sensation or symptom itself, as well as the thoughts and emotions that may be associated with the symptom, allows the individual to recognize the sensation itself. In turn, individuals may become less attached to the associated thoughts and emotions, which enables more opportunities for flexible responding to stress and somatic symptoms. With practice, the sensation itself becomes more manageable and the child becomes less limited by it. When considering integrating mindfulness techniques into the care of your patients, it is essential to identify mindfulness instructors with excellent training and experience and ongoing support for their practice. In the right hands, mindfulness meditation instruction has extraordinary potential for benefit.

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Adapting Yoga for Children and Adolescents with Functional Disorders

25

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Abstract

Yoga practice is intended to reduce suffering and encourage optimal functioning through self-awareness and self-transformation. A basic assumption underlying the practice of yoga is that the mind and the body are mutually influential and the practices of yoga are intended to simultaneously and holistically effect change in the whole person. The general aim of yoga practice is to bring the body and mind into a balance of stability and ease at increasingly more subtly perceptive levels. In this chapter, the importance of matching the practice to the person in order to increase motivation and maximize potential benefit is emphasized. Information is presented about differences in yoga styles and classes and the ways that yoga techniques can be applied and adapted to the needs of the individual and to children. Evidence for the effectiveness of yoga as an intervention for functional disorders is reviewed. Case studies are presented demonstrating use of yoga techniques with adolescents diagnosed with irritable bowel syndrome and fibromyalgia. Videos, images, and instructional text are provided.

Keywords

Yoga • Asana • Breathing • Meditation • IBS • Fibromyalgia

Yoga Philosophy

Yoga is an ancient methodology for the relief of suffering and optimization of human potential. Over the estimated 5,000 years that yoga has been practiced, human beings have utilized yoga principles and practices as a means to increase their capacity to live healthy, happy, and meaningful lives. The core principles of yoga are said to be *Sanatana Dharma*, universal truths that are applicable to all people in all times regardless of age, gender, culture, and religion [1]. Across time, the particular techniques and associated practices of yoga have adapted to the changing cultural contexts in which they have been practiced; however, the core philosophical tenents have remained a constant foundation and guide.

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Yoga philosophy posits that individuals have the ability to lead joyful lives free from fear and suffering and that this is the true state of human consciousness (in Sanskrit *Satchitananda*). Consciousness (*Purusha*) pervades, animates, and organizes expression in all life. Human consciousness is self-reflective; therefore humans are able to remember experiences, to compare across time, and to differentiate between pain and pleasure. Humans have an innate preference for pleasure and drive toward growth and development. These can be thwarted and shaped by life circumstance so that they are pursued in less effective or beneficial ways, but the drive to seek them persists. The yoga sages assert that consciously or unconsciously the individual remembers that freedom from fear and joy without pain are possible. Consequently, the individual longs for and seeks through more or less constructive means to experience that state. *Satchitananda* is the state of an individual who consciously remembers and can live in bliss consciousness while in the world.

Yoga practices were developed as a means to enable individuals to have their own experiences of what the yoga philosophy asserts is optimally possible for human beings to know, understand, feel, and do. Yoga practice provides individuals with a combination of knowledge (mind) and state-changing techniques (body–mind) to effectively expand their conscious awareness and build their capacity for skillfully influencing the direction of change in their lives. Step-by-step the practice affords new experiences that expand and shift perspective, thus giving a taste of what is possible while building an individual's trust in his own self-efficacy to achieve it. Personal experience, *darsana*, or "direct vision" is necessary to catalyze the innate human potential to grow beyond current limits of ability and perception and in the process to transform perception and increase ability. Yoga practices provide the methods by which an individual can volitionally, repetitively, and reliably alter his/her physical, physiological, and mental/emotional states in tandem. The word yoga comes from the Sanskrit root word *yug* which means to yoke. Just as two oxen are yoked together and the pace of one affects the smoothness of their pace together, the physical structure and physiological systems of the body, the mind, and the consciousness that animates the whole are yoked together. Disruption in one causes disruption in the other. A lack of coherence leads to less effective action, compensation, and greater strain on various parts of the system. Left uncorrected, this dysregulation leads to system breakdown in whichever areas are under greatest strain or most vulnerable. Yoga practices are designed to foster improved health and balance, so the person is able to experience life with less restriction/greater freedom and less pain and suffering/greater joy.

The practical application of yoga philosophy is manifested in the step-by-step conscious choices an individual makes to improve the quality of his/her life. Quality of life is improved by acting in ways that create greater stability and equilibrium, greater flexibility and freedom, and greater relief from suffering and joy in both the mind and body. Yoga practices provide an individual with the empowering experience that he or she can positively affect the way he or she feels with reliable predictability by doing simple actions such as focusing attention, regulating the flow of the breath, and initiating pain-free movement in the body. These are vital experiences to enhance the self-efficacy of a child whose identity, perceptions of the world, and the way he or she interacts with the environment are being crafted. They are even more crucial experiences for a child with a functional disorder who may lack the skills and confidence to meaningfully improve his condition.

The teachings of yoga were passed down from teacher to student through oral tradition before they were written down. The earliest texts that form the written record of yoga philosophy and practice are called the *Veda*s, which translates "to know," written approximately 3000–1500 BCE, and the *Upanisad*-s, which means "sitting down near [a teacher]," composed 1500–700 BCE [2]. As the names convey, increasing knowledge and understanding are central quests, and transmission of wisdom is done in the context of an established relationship between teacher and student.

The first mention of the word "yoga" appears in the Katha Upanishad dated around 400 BCE [1, 3]. The *Katha Upanisad* is written in the

form of a teacher–student dialogue, as are many of the *Upanisads*. The teaching given is that pure consciousness is eternal and unchanging and does not die with the body. Pure consciousness is the same animating force that humans conceive of in different personal and universal forms: the soul, the Self, the order of the universe, and the Divine. Having direct experience of this transcendent unitary reality, and identifying with it rather than the fleeting pleasures and pains of the body and world, is revealed as the method to overcome all grief, fear, and the hunger for temporary pleasures. Developing a still, calm mind with senses and attention focused inward is a preparatory step, and this state of mind "is called yoga" (p. 133) [3]. The pathway to inner peace is meditation, repetitively redirecting the mind inward and focusing it on a single object of attention so that the clamor of many thoughts becomes absorbed in the one.

The Bhagavad Gita, dated variously between 200 BCE and 200 AD and also written as a teacher–student dialogue, delineates the three paths of yoga (1) the yoga of action (*karma yoga*), (2) the yoga of knowledge (*jnana*), and (3) the yoga of devotion (*bhakti*) [4]. The teacher asserts that the best path is the one that enables the specific individual to fulfill his life purpose. *Karma* yoga is the path for most people as most are oriented to action in the external world, such as raising a family and doing a job. Individuals who are on this path are exhorted to act "without regards to the fruit of your actions," and yoga is defined as "skill in action" and "equanimity" [4]. Acting without regard to the fruits means to do what is the person's responsibility and duty without being deterred or influenced by expectations of reward, praise, or criticism. In other words, the actions and opinions of others should not sway a person from doing what is his to do and is right to do. *Jnana* yoga is for introspective and intellectual individuals who seek to gain understanding through rigorous observation and reflection. This is said to be the hardest path because these individuals must renounce many of the pleasures of everyday life in order to divert their attention internally. *Bhakti* is the path for individuals of a devotional nature whose primary actions are to serve others. Despite delineating the paths as separate, like all things in yoga, the qualities are interrelated, and the yogi is entreated to embody the qualities of all three paths in order to live a full and complete life. For example, even for the intellectual, some actions must be performed, so they should be done without regard to the consequences and with attention to the needs and service of all beings. See Table 25.1 for additional information regarding the paths of yoga.

Table 25.1 Paths of yoga

Four paths of yoga	
Bhakti yoga	Path of devotion
Jnana yoga	Path of knowledge
Karma yoga	Path of action
Ashtanga yoga/Raja yoga	Eight-limbed path/royal path
Eight limbs of Ashtanga/Raja yoga path	
Five Yamas: Ahimsa, Satya, Asteya, Brahmacharya, Aparigraha	Five restraints: nonviolence; truthfulness; non-stealing; sexual moderation; lack of greed/non-possessiveness
Five Niyamas: Saucha, Santosha, Tapas, Svadhyaya, Ishvarapranidhana	Five observances are mental disciplines: physical and mental cleanliness/purity; contentment; discipline/austerities to make the body strong and mind impervious to changes in the body; self-awareness/self-study; offering oneself in service to the Divine, surrendering self-will
Asana	Means "seat" but is used to connote physical postures
Pranayama	Regulation of the breath, in particular to slow breath to the point of consciously creating "pauses" or "gaps" between the need to inhale and exhale
Pratyahara	Withdrawal of the senses, drawing the senses inward
Dharana	Focused concentration on one object of attention
Dhyana	Sustained one-pointed concentration
Samadhi	Consciousness merges with the object so that differentiation between self and object disappears and an experience of oneness/complete absorption in the object is all that remains

The Yoga Sutras of Patanjali, also dated between 200 BCE and 200 AD, is a collection of 196 aphorisms (*sutras*) [5]. The Yoga Sutras provide guidance about symptoms of disease, common obstacles to practice, ways to overcome the obstacles, signs of mastery, and successive stages of new capacities that may develop. This information is offered to give anticipatory guidance, normalize experience, strengthen the will, and inspire faith to continue the practices that lead to self-transformation.

The Yoga Sutras describe the fourth path of yoga called the eight-limbed (*ashtanga*) and also the royal (*raja*) path (Table 25.1). Each of the eight limbs is preparatory for the next but they are nonlinear so the student may progress in several simultaneously. Changes in the way individuals interact with others in the external world are recommended first so that the surrounding environment is harmonious and the amount of external and internal conflict and stress are minimized. Cultivation of a strong and healthy body is introduced next in order to decrease the mental distractions that arise when the body is in pain or compromised by illness. When the body is stable and at ease, the mind can attune to more subtle internal sensations including the breath. Breathing practices are used to regulate the autonomic nervous system thereby bringing the physiological systems of the body under more conscious control and promoting optimal functioning in them. Drawing the senses inward is another step away from external distraction and toward inner self-observation. The first five limbs are preparatory to meditation, which begins with the sixth limb by focusing attention on one object. Greater ability to sustain attention on one object, and then complete union with this object, results from the elimination of mental distractions and the ability to direct the mind at will to be completely absorbed in the single focus of attention. This final step, union with the object, is the method for achieving the radical shift in perspective that leads to experience of the true Self and pure consciousness. Self-realization, also called enlightenment or *Samadhi*, leads to the unitary realization of the interconnection of all beings, feelings of bliss, compassion, and complete understanding. The eight limbs provide techniques to shift the person into a sustained unified internal state through which the new perspective becomes possible and the optimal capacity of the individual is achieved.

The first texts to detail and emphasize the physical practice of yoga were written between 600 AD and 1500 AD. The Hatha Yoga Pradipika, which translates as Light or Illumination on Hatha Yoga, was compiled during this period [6]. The word Hatha is a combination of *ha*, which indicates *prana*, the vital life force, and *tha*, which is *manas shakti* or mental energy [6]. Hatha yoga practices aim to increasingly facilitate balance and integration between these two energies. Hatha yoga begins with purifying, detoxifying, strengthening, and bringing the systems of the body into healthy and effective functioning. The basic premise is that when the body is not functioning well, neither does the mind, and it is easier to restore function to the body than the mind. Discipline and self-regulation are introduced through postures and breathing practices instead of first attempting to regulate thoughts and feelings as the four paths previously described tend to do. It is posited that the physiological shifts achieved through body and breath practice naturally facilitate concomitant mental shifts in attitude and affect that facilitate greater readiness and equilibrium when the attention is turned inward.

The Hatha Yoga Pradipika describes *asanas* and their reputed benefits, practices for cleansing and purifying, regulating the breath, and cultivating, directing, and containing vital energy [6]. These more advanced practices are incorporated into practice as the individual becomes capable of increasingly refined internal attention and affective and behavioral regulation. See Table 25.2 for definitions of yoga practices and Table 25.3 for descriptions of holistic kosha model and energies.

Modern Yoga Practice

The number of individuals practicing yoga is steadily growing. In 1998, national surveys estimated that 3.8 % (7.4 million) American adults

Table 25.2 Sanskrit terms for yoga practices

Yoga practices	
Abhyasa	Yoga practice
Bandha-s	Energy locks, Mulabandha = pelvic floor, Uddiyanabandha = navel center, Jalandharabandha = chin lock
Brahmana	Practices with an energizing or increasing effect
Hatha yoga	Ha represents prana, life force; Tha represents manas shakti, mental energy
Japa	Silent mantra repetition
Kirtan	Chanting mantras
Kriya-s	Practices for cleansing, purifying, and detoxifying
Langhana	Practices with a calming or reducing effect
Mudra-s	Gesture made with the whole or part of the body, e.g., hand that embodies an attitude or creates a seal
Nadi shodhana	Alternate nostril breathing; breathing in one nostril and out the other
Pawanmuktasana	"Joint freeing" systematic gentle movements of all the joints of the body
Pratipaksha bhavana	Acting opposite
Sama vritti	Even duration of inhalation and exhalation
Sankalpa	Setting an intention
Sat Nam	Mantra meaning I Am Truth. May also be separated into parts and chanted as Sa Ta Na Ma
Savasana	Supine relaxation posture, "corpse pose"
Surya namaskar	"Sun salute," fixed sequence of poses
Vairagya	Letting go of attachment
Yoga	To yoke together, union
Yoga nidra	"Yogic sleep," multipart guided relaxation

Table 25.3 Holistic kosha model and energies

Holistic kosha model	
Five kosha-s	Sheaths/layers of the self and Self
Annamaya kosha	Physical structure of the body
Pranamaya kosha	Energy body; prana = life force, includes all physiological systems, e.g., cardiorespiratory, immunological, digestive, endocrine, and autonomic nervous system
Manomaya kosha	Mental/emotional body; includes thoughts, feelings, attitudes, beliefs, memories
Vijnamaya kosha	Intuitive body; "gut" or "heart" or "wise mind" way of knowing what is right for you, inner teacher, higher self
Anandamaya kosha	Bliss body; connection to source, Divine, universal intelligence
Manas shakti	Mind/mental energy
Prana shakti	Vital energy; life force

had practiced yoga in the previous year [7, 8]. By 2012, a market survey conducted at the behest of Yoga Journal magazine reported a rise to 8.7 % (20.4 million) adult Americans practiced yoga in the previous year [9, 10]. Over half (53.6 %) of respondents in an East Coast University survey published in 2012 endorsed having practiced yoga [11]. The only survey estimating prevalence use for youth reported in 2007 that 1.5 million children and adolescents had tried yoga [12].

The demographics of yoga participants are similar across the surveys. Yoga practitioners are more likely to be female (range reported 68–82 %), Caucasian, college educated, and urban dwellers, who are also likely to use other complementary and alternative medicine (CAM) approaches [7–11]. More than half of the respondents endorsed using yoga to improve and maintain health and well-being (range 58–64 %) [7, 8]. Estimates varied on the number of individuals who used yoga to treat a specific health condition (16–48 %) [7, 8]. The four most commonly reported conditions for which yoga was used as a treatment were (1) musculoskeletal (arthritis, rheumatoid arthritis, gout, lupus, fibromyalgia, and joint pain), (2) severe sprains, (3) mental health (depression, anxiety), and (4) asthma [7]. The individuals who used yoga to treat a condition described it as very helpful: 83 % for mental health, 76 % for musculoskeletal conditions, and 90 % for wellness [7, 8].

The modern practice of yoga is heavily influenced by the physical focus of hatha yoga. In the common parlance, "doing yoga" is equated with doing poses. The different styles and schools of

yoga vary in the degree to which they focus primarily on asana practice or include the other five common elements: breathing practices, energy regulation, relaxation, meditation, and yoga philosophy. Coordinating breath with movement is routine in most yoga classes, but other forms of breath regulation or meditation may or may not be included. Most yoga classes end with a period of rest in a supine relaxation posture (*savasana*). Energy and attention regulation practices described in the Hatha Yoga Pradipika are often included in advanced classes and workshops, and they are a core component of Kundalini yoga. Yoga concepts and philosophy are often addressed at the beginning to set the intention of the class and may be woven through in the instructional language of the class to cultivate a particular attitude or focus of attention. Beyond the core elements, the principle variations in yoga styles/classes include:

1. *Using a fixed or varied sequence of poses and practices*. The degree to which teachers instruct predetermined sequences varies from being extremely fixed, meaning the same poses are taught in the same order each class, e.g., Bikram, and predominately prescribed, e.g., Ashtanga series, to teacher's choice. Fixed sequence classes offer the benefits of predictability, opportunity for mastery through repetition, and easy comparison day to day about changes in state and ability. These factors can make fixed sequence classes appealing to beginners and high achievers. However, even in the more variable classes, teachers tend to incorporate familiar elements and orders so these benefits can also be attained by regular class attendance with the same teacher. Each element in yoga practice is intended to produce certain effects. The art and science of sequencing is choosing which techniques to include and in what order to place them to produce the intended effects. The way the practices are ordered impacts the degree to which the student is mentally and physically prepared for stronger and more challenging elements, which may serve to decrease the risks and increase the benefits of individual elements. Counterposing is necessary to neutralize the stronger practices and to restore ease and balance.
2. *Variation from gentle to vigorous and challenging*. Yoga classes taught to young and fit individuals tend to be more vigorous and challenging because the students often have a need to discharge rather than accumulate energy and their minds and bodies are primed for acquisition of new abilities. High-energy and anxious students also benefit from vigorous classes that discharge energy and focus the mind. Some other individuals are better served by slow, gentle, and energy-building practices. For the majority of individuals with functional disorders, gentle forms of yoga are recommended at least during acutely symptomatic periods.
3. *Fast- or slow-moving transitions between poses or long held poses*. Classes in which movement flows from pose to pose, such as Vinyasa Flow, tend to be fast moving. At the other end of the spectrum, in Restorative and Yin yoga classes, students remain in a pose for 5–20 min. An interim technique is to combine moving in and out of poses with staying in a single pose for approximately five breaths. The techniques all produce different muscular and cardiovascular effects and have quite opposite effects in terms of stimulating the sympathetic versus the parasympathetic nervous system into dominance.
4. *Degree of use of props* (*blankets, bolsters, blocks, straps*). Props are used to support the body when it is held still in poses and to enable individuals to move into a pose that they would otherwise have difficulty achieving or benefiting as fully from. Classes focused on alignment and therapeutics such as Iyengar classes make extensive use of props. Restorative yoga classes in which the goal is to achieve deep, prolonged rest also use props.
5. *The temperature of the room may be moderate or greatly heated*. At the extreme end, the room temperature in Bikram classes is heated to 105 F in order to relax muscles and promote detoxification through sweating. "Hot hatha" classes raise the temperature to 80–90 F. These temperatures combined with movement

can induce dizziness and nausea in some individuals and may exacerbate symptoms in persons with heat-sensitive disorders such as multiple sclerosis. Others may experience difficulty if the outside temperature after leaving class is considerably different. Otherwise, yoga classrooms tend to maintain the heat between 70 and 80 F.

The key factor in choosing a yoga class is goodness of fit. The practical aspects of location, time of day, and cost need to be balanced with more personal and relational considerations. It is important to feel trust, safety, and inspiration with the teacher; to have an appropriate fitness capacity for the activity level of the class; and to feel benefit from participation. When describing the paths of yoga, the point is emphasized frequently that matching the method to the interests and abilities of students will increase the likelihood that he or she will adhere to the practice. Motivation is a vital consideration because yoga, unlike many other healthcare interventions, is a self-help, lifestyle change strategy. Since the practice must be repeated and maintained over time for the benefits to become ingrained, finding a practice the person enjoys and can persist with is essential.

Adaptation of Yoga for Health and Children

Group yoga classes have the benefit of shared social interaction, reduced cost, and increased access for a larger number of people, but personalized one-on-one instruction geared to the specific level and interests of the student is forfeited. Open community classes are intended to provide general wellness benefits to the mind and body. However, as noted above, a growing number of individuals are participating in yoga classes with diagnosed disorders and are seeking assistance with specific conditions. Yoga teachers are encouraged to provide students with modifications to poses at different levels of difficulty to accommodate the broad needs of the class, but differences in the style of yoga being instructed and the skill and experience level of the teacher will vary the degree to which attention is paid to individual needs.

Yoga therapy is emerging as a profession in response to the growing number of individuals who are seeking to obtain health and wellness benefits from yoga. Drawing broadly on the rich repertoire of yoga philosophy and practice, yoga therapists are trained to adapt techniques to the specific and holistic needs of the individual and to design personalized routines for ongoing home practice [13, 14]. Yoga therapists are experienced yoga teachers who have undergone additional specialized training to work more broadly and in depth with their clients than yoga teacher training prepares them to do. Some yoga therapists are also western- and eastern-trained health providers, and they are intentionally seeking to bridge yoga with other health-care approaches. The International Association for Yoga Therapists (IAYT) has formulated minimum training standards for yoga therapy training programs. For more information, see resources in the Appendix to this book.

Additional specialized training is also recommended for yoga teachers who wish to work with children and adolescents. Yoga teacher training courses generally focus on teaching yoga to adults, and there are developmental considerations that must be taken into account when teaching children. Training is available for teaching pregnant women, mothers and infants, mothers and toddlers, school-age children, and adolescents. The instructional language must be adapted to the attention span, visual-motor coordination capacity, and cognitive level of the child [15]. The poses and breathing practices are still used, but an attitude of playfulness is usually incorporated into the language and the names used for the practices. For example, in the Yoga Ed curriculum, the poses are called by their nature names, e.g., mountain, bear, and rock; breathing practices include snake, bunny, and dog; and meditation is called "time in" [16]. Other elements may include singing songs, integrating the poses into stories, and playing games that incorporate yoga techniques. Yoga philosophy is imparted through the attitude and emphasis on building competence and the confidence to go

with it, creativity and resourcefulness, feelings of peacefulness and joy, and trust in oneself in the context of inclusive community. For example, the Rainbow Song in the Yoga Ed curriculum teaches the qualities of the seven *chakras*, areas of energy concentration, as follows [16]:

> Red, I'm strong,
> Orange, I'm joyful,
> Yellow, I know I can,
> Green, I'm loving,
> Blue, I tell the truth,
> Indigo, I know,
> Violet, Understanding.
> I'm a Rainbow!

Swami Saraswati of the Bihar School in India, who wrote the classic book *Yoga Education for Children*, stresses the importance of introducing yoga practice to children early in their lives so that they have well-established tools for managing the physical and mental changes of puberty [17]. He recommends initiating boys and girls into practice at 8 years old, as was traditional in certain segments of Indian society, with an active physical practice of fixed sequence poses (*surya namaskar*), a balancing breathing practice (*nadi shodhana*), and a mantra (*Gayatri*). "Yoga," he writes, "is a form of complete education that can be used with all children because it develops physical stamina, emotional stability, and intellectual and creative talents. It is a unified system for developing the balanced, total personality of the child" [17]. In the USA, yoga classes are increasingly being incorporated into the school curriculum in order to provide children with mindful movement, enhance concentration and attention, and build capacity for emotion regulation. See the Appendix for additional resources.

Empirical Evidence for Yoga

The majority of yoga studies are conducted with adults. In addition, relative to other conditions of interest, the study of yoga with medical conditions in children is profoundly underrepresented in the literature. Despite the limited number of trials, there are three meta-analytic reviews evaluating the effectiveness of yoga as an intervention with children and young adults [15, 18, 19]. Overall findings are of positive benefit in both physical and mental/emotional domains, but the research methodology needs improvement to increase confidence in the results.

The National Institutes of Health's National Center for Complementary and Alternative Medicine (NIH NNCAM) summarized the scientific evidence for physical benefit from yoga across all age groups to include improvement in function; overall physical fitness, strength, and flexibility; lowering of heart rate and blood pressure; and reduction in back pain. The evidence for improvement in asthma and arthritis was inconclusive [20]. Evidence supportive of mental/emotional benefit includes improvement in quality of life and reductions in stress, anxiety, depression, and insomnia [20]. These findings are consistent with the avowed intentions of yoga practice to promote enhanced well-being and quality of life.

Biopsychosocial models used to explain the effects of yoga posit multiple system involvement, as would be expected for an intervention that aims to be holistic. The systems most often identified as positively impacted by yoga closely match the systems that are described as dysregulated in many functional disorders, including irritable bowel syndrome (IBS) and fibromyalgia [21–27]. In the physiological realm, it is proposed that yoga helps to regulate the endocrine system including hypothalamic–pituitary–adrenal (HPA) axis reactivity, cortisol activation patterns, and autonomic nervous system (ANS) regulation [23, 28]. Yoga is postulated to support ANS balancing by reducing sympathetic overactivation and increasing parasympathetic activity through stimulation of the vagus nerve during asana and breathing practices, leading to increased vagal tone and heart rate variability (HRV) [26, 29].

Yoga is hypothesized to reduce stress-related release of proinflammatory cytokines. Novice yoga practitioners have been shown to have higher average interleukin-6 (IL-6) levels, greater likelihood of having detectable high-sensitivity C-reactive protein (hsCRP) levels, and to release more LPS-stimulated IL-6 in response to intervention stressors than experienced practitioners

[30]. One of the aims of yoga is to achieve and maintain a state of equilibrium, which may serve a protective function to reduce stress reactivity and burden.

Specifically in terms of IBS, yoga asana is additionally proposed to have a direct peripheral effect on gut muscles through pressure changes, which may stimulate visceral afferents in the enteric nervous system [27].

Psychological effects from yoga practice include reductions in anxiety and depression, which are commonly comorbid in individuals with functional disorders, and improvement in coping and self-efficacy, which are needed by individuals with functional disorders [23, 25, 31–34]. Low GABA neurotransmitter levels have been linked to depression, and they have been shown to rise in response to yoga practice, suggesting increasing GABA levels may be one physiological mechanism through which yoga fosters positive psychological state changes [25, 29, 30, 35]. Two studies conducted in schools with adolescents found that while yoga did not improve mood in the participants, it served a protective function by helping to maintain mood levels during periods when the control group's subjects reported significant worsening of mood [36, 37]. In addition, the yoga participants reported increases in resilience and decreases in anger and fatigue, in contrast to the control group who reported the opposite effects [36]. In a sample of college students, yoga was shown to significantly reduce cortisol levels, perceived stress, and negative affect immediately following a single 90-min class [38].

Irritable Bowel Syndrome

Only two published reports have evaluated yoga's effectiveness as an intervention for IBS: one with 22 adult males (9 in the yoga group, 13 in the control group) in India with diarrhea-predominant IBS [39] and the other with 25 adolescent females (all received the intervention in a wait list control design) in Canada with mixed-type IBS [40]. In both cases the participants were asked to practice on a daily basis: the males twice a day for 2 months and the females 10 min once a day for 4 weeks. Both interventions included backward and forward bending asanas and side bending or twisting poses to alternately contract and release the musculature of the abdomen. In all cases the male cohort performed more challenging versions of the poses, which are presumed to be more activating of the sympathetic nervous system. The males utilized an alternate nostril breathing practice at the end which is again presumed to be more stimulating (*surya bhedana*), whereas the females began the practice with deep, slow parasympathetic activating breathing while prone on their abdomens. In the male study, the yoga participants reported improvements in bowel symptoms equivalent to the control group benefit from loperamide. The yoga group showed greater parasympathetic activation on a deep breathing ECG test and reduction in state anxiety than the controls. At the end of the adolescent female study, there were a significant reduction in gastrointestinal symptoms and a trend ($P=0.10$) toward lower levels of emotion-focused avoidance in comparison with a wait list control group. In a post-intervention qualitative assessment, the adolescents reported they found the yoga practice helpful (mean = 7.2 out of 10), very enjoyable (8.2), and easy to do (9.2), and they planned to continue using it (24 of 25 respondents).

Fibromyalgia

Two recent studies of adult women diagnosed with fibromyalgia found that a combination of using gentle mindful movements, discussing yoga principles, giving specific information about how to practice safely when experiencing pain, and fostering a "non-judgmental, compassionate and accepting approach to practice" led to significant improvements in physical and mental symptoms. These studies demonstrated reductions in pain, fatigue, stiffness, tenderness, and poor balance, as well as lessening of depression and anxiety, improved memory [41], decreases in pain catastrophizing, and increases in mindfulness [42]. Curtis et al. hypothesized that the instructions to move mindfully with acceptance

and without judgment contributed to participants having present moment interoceptive (mind–body awareness) experiences that countered their habitual avoidance of movement due to fears of experiencing pain [42]. Interestingly, a yoga program developed specifically for psychologically traumatized youth follows very closely the core values and components espoused in the fibromyalgia studies, and the program describes its model to teens as a process to "reclaim your body" [43].

Case Studies

Case 1: Fibromyalgia

Katy, age 16 and diagnosed with fibromyalgia, was referred by a nurse in the pain clinic to a hospital-based yoga research study for adolescents with mental and physical health concerns.

Katy had the hallmark complaints of pain in her muscles and joints that shifted in location and intensity, feelings of fatigue that waxed and waned, and symptom flare-ups that seemed to come on randomly. Katy defined herself by her diagnosis. She felt that fibromyalgia dictated the terms of her life, and nothing she did predictably resulted in relief or improvement.

In the first few weeks, Katy frequently interjected that movement "hurts." In response, Katy was encouraged to pay even closer attention to her perceptions to determine if she felt sensation, the effect of her movements and muscles working, or if she felt pain. Katy had avoided movement for so long that when she perceived sensation, it was pain to be avoided. Katy was consistently and calmly guided to move slightly out of the pose until she found the point where there was sensation but not pain, rather than reactively pulling all the way out of the pose as she had initially done. This gradual movement with awareness allowed Katy to begin to re-titrate her pain perception. She was also offered modifications to the poses that enabled her to get the benefits of the pose without pain, thus teaching her to think flexibly about alternatives.

Just as with movement, mental attitude affects the ability to remain still and shapes the experience of stillness as peaceful or agitating. Katy was initially unable to rest comfortably in *savasana*, the supine relaxation pose that is the final posture in most yoga classes. To cultivate her mental and physical abilities to let go of tension and relax in the pose, Katy was guided through a *pranayama* and visualization practice called point-to-point breathing [44].

Point-to-point breathing is intended to unite the mind and breath by moving awareness in tandem with the breath through various points in the still, relaxed body. The breath flows in and out as awareness moves up and down the body. The instructions are to let awareness flow up and out of the body like water washing through bringing a sensation of spaciousness and release. This practice was chosen for Katy to focus her attention in her body while giving her a technique to let go of habitually held tension in her body.

The following are available:

1. Videos of an 18-year-old female being guided in point-to-point breathing
   (a) Video 25.1 Sitting preparation
   (b) Video 25.2 Relaxation preparation
   (c) Video 25.3 Point-to-point breathing practice
   (d) Video 25.4 Moving back into awareness
2. Instructions for point-to-point breathing
   (a) Table 25.4 Introduction
   (b) Table 25.5 Relaxation preparation
   (c) Table 25.6 Practice instructions
   (d) Table 25.7 Moving back into awareness

Questions

1. Which element describes mindful yoga practice?
   (a) Moving with awareness, purposefully attuning to the sensory effects of movement
   (b) Tuning out and trying to distract the mind with enjoyable stimuli
   (c) Blocking out all thoughts to quiet the mind
   (d) Powering through with a no pain, no gain attitude
   (e) Playing it safe, not doing anything that may cause pain
2. Which is NOT one of the potential benefits of adapting yoga poses to the individual?
   (a) Developing mental flexibility
   (b) Learning to think creatively

Table 25.4 Introduction to point-to-point breathing practice

Point-to-point breathing is a practice of moving awareness through your body in harmony with your breath. As your breath moves, your awareness moves, and the two move together up and down your body from the soles of your feet to the crown of your head and seven points in between
The points where you will focus awareness are: 1. The soles of your feet 2. Your Ankles 3. Knees 4. Hips and pelvic bowl 5. Navel center 6. Heart and the center of your chest 7. Throat 8. Forehead space between your eyebrows 9. Crown of the head—at the center and top of your head
You may do this practice lying on your back on the floor or in your bed. Before you begin the practice, gather together a yoga mat if you have one, a blanket to keep the body warm when your temperature cools during relaxation, a thin pillow to support your neck, and an eye pillow to relax the eye muscles and decrease sensory stimulation from light and sight
During the practice, you will move your awareness between the points in your body as you breathe in and out with a deep, slow breath taking four breaths at each point. The regular rhythm of the breath guides your mind and body into balance, which is soothing. The long deep breaths bring oxygen to the tissues of the body, which is revitalizing. Sustaining this rhythm over a period of time is deeply nurturing to your mind and body allowing them both to relax and be refreshed at the same time
We will end the practice with a short period of resting in stillness. Then I will guide you back into movement and back into your day
The more regularly and often that you do this practice, the easier you will find it to settle into the rhythm and reap the benefits of the practice. It may take a few tries to become comfortable with the practice, or you may find that you take to it right away. In either case, you will find that some days it is easier than others. Some days your mind will be more restless or distracted, and some days your body will be in more pain. Whatever arises in your body or mind, stick with the practice directing your mind back again and again to the rhythm of your breath moving with awareness through your body together

(c) Building confidence to listen and respond to the body
(d) Strengthening will power
(e) Building courage to do something slightly different than others

Table 25.5 Getting settled in relaxation pose

To begin the practice, lie down on the floor or your bed
• Open your arms wide by your sides a comfortable distance away from your body with the palms turned up
• Extend your legs straight on the floor with the toes and feet relaxed away from each other
• Support your neck and head with a pillow that allows your head to rest in a neutral position
• You may want to place a bolster or rolled blanket beneath your knees to relieve pressure in your lower back
• During the practice your body temperature may drop, so cover yourself with a light blanket
• Once you are comfortable, if you have an eye pillow, place the pillow over your eyes
Let your body relax. Feel the floor or bed beneath your body, and let your body soften into the support below. With each exhale, feel your body become heavier as it softens into the support beneath it
• Pause 15–30 s
Bring your attention to the space surrounding your body. Feel the space above, below, and beside your body. Become aware of the feel of your skin and the place where the space gently touches your skin. Allow your body to relax and soften into the space that surrounds you
• Pause 15–30 s

3. In which case would it be recommended to move gently to the point before pain?
   (a) In cases of acute injuries
   (b) When stabilization is required
   (c) When there are pain avoidant symptoms of fibromyalgia
   (d) When there is acute inflammation
   (e) When the pain is not as bad as the person thinks

Answers

1. (a): Mindful yoga involves being present with sensations as they arise.
2. (d): Strengthening the power of will and determination are important qualities in sticking to a regular yoga practice, but pushing through a yoga practice that is not beneficial is not appropriate adaptation of the practice to the person.
3. (c): Fibromyalgia is neither a case of acute injury or inflammation, so restoring confidence in skillful movement is actively modeled and encouraged.

Table 25.6 Point-to-point breathing practice instructions

(NOTE: Observe the client's breath and time your instructions to match the length of inhale and exhale and to match when he or she has completed four breaths in each location. Use fewer words during shorter distances between soles of the feet and the named body part and more words for longer distances to help gradually lengthen then shorten the rate of breath. Give instruction and guidance for the first breath, then silence for the next three breaths. Alternate using visualization guiding words such as "feel spacious," "wash out," "release, let go," "flow," and "fill the space with light and ease." Prompt to take three more breaths.)
• We will begin the practice now. Simply direct your attention to the part of your body I name, and take four breaths up and down from that point. Begin by bringing your attention to the soles of your feet
• Focus your attention on the space touching the soles of your feet. On your next breath in, draw your awareness up from your feet to your ankles, and as you exhale, feel your awareness flow back out the soles of your feet. Take three more breaths with your awareness traveling upward with your inhale from the soles to your ankles, and as you exhale, awareness flows back down through the soles of your feet
• Next breath in your awareness rises up to your knees. As you exhale your breath and awareness flow back out your feet. Three more breaths from your feet to your knees. As you inhale feel your knees become spacious. As you exhale feel your awareness flow down your legs and out your feet
• Hips and pelvic bowl
• Navel center
• Heart
• Throat
• Forehead, the space between your eyebrows
• Breathe in, awareness flowing all the way up to the crown of you head. Feel the breath rise up through your legs, body, and mind to the crown of your head. On your exhale, feel the breath flow back out through your mind, body, legs, and feet. Three more full body breaths. Feel the breath flow uninterrupted from the soles of your feet to your crown and the crown of your head to the soles of your feet
Rest your awareness in your body. Feel yourself as spacious, at ease.
• 1- to 5- or 10-min pause in silence

Table 25.7 Moving back into awareness

Bring your awareness back to your breath and slowly begin to deepen it. With each breath in, feel your breath becoming more full and complete. Feel the breath moving in your belly and chest
• Observe and count five breaths
Gently begin to move your body starting with toes and ankles
• Wiggle your toes. Rotate your ankles in circles. Now make ankle circles in the other direction
• Wiggle your fingers. Move them in and out of your palm making a gentle fist and letting it go. Now make circles with your wrists. First one way, then the other way
• Let your hands and feet rest and gently turn your head side to side
• Stretch your hands over your head, stretch from fingers to toes on inhale, and on exhale release body and breath together. Two more times
• Roll over to your right side and take a few breaths.
• Use your hands to press against the floor or the bed to gently push yourself up into a comfortable seated position
• Take a moment to scan your body, toes to head, and notice how you feel now
• Notice the quality of your breath
• The quality of your mind
• Take a moment to thank yourself for taking the time to do your practice and take care of yourself
• You can now move back into your day

Case 2: Irritable Bowel Syndrome

Charlotte was 16 years old when she started private yoga therapy. Her physician diagnosed her with irritable bowel syndrome (IBS) and her psychiatrist with obsessive-compulsive disorder. Charlotte's physician recommended yoga to her, but she felt too shy and uncomfortable in her body to join a group class.

Charlotte's IBS was the bloating and constipation type. Charlotte felt out of control of her own body, and she was both frustrated and embarrassed that she could not better manage herself. Charlotte held herself to a high standard of performance wanting to do the poses "perfectly." She frequently asked if she was doing things "right" and often commented that she was doing things "wrong." It took a period of time with much repetition to shift Charlotte's focus from the external look of the pose to its internal experience.

Charlotte was guided through a short series of poses that were intended to gently stimulate coordinated colonic motility and simultaneously facilitate a relaxation response. Charlotte was encouraged to practice the same poses at home on most days. She was encouraged to experiment to find out if she benefited more from doing them in the morning, afternoon after school, or at bedtime.

The majority of the poses included forward bending (flexion) because forward bending tends to have a calming effect. In each posture Charlotte moved in and out of the pose with attention to the inhalation and exhalation of her breath. To gently and rhythmically compress the abdomen, her legs were drawn into the chest symmetrically (both knees at the same time) and asymmetrically (one knee at a time). Coordinating gentle abdominal compression with a slow balanced inhale and exhale is used to trigger a shift to parasympathetic dominance and to stimulate syncopated colonic motility. A supine twist was also included because twists engage the muscles of the abdomen to contract and release and correspondingly seem to foster a mental attitude of letting go upon release of the twist. Charlotte experienced a reduction in bloating and lessening of constipation fairly quickly after a regular practice was established. This tangible benefit increased her motivation and confidence that using yoga she could improve her condition.

Due to her perfectionism, Charlotte found school stressful. She was open to trying something to help her feel more calm and focused when she could not do a yoga pose, so she was taught an unobtrusive *mudra* and *mantra* practice she could do at any time. The practice is to silently repeat the mantra Sa Ta Na Ma while touching her thumb sequentially to each of her four fingers: Sa thumb to forefinger, Ta thumb to middle finger, Na thumb to ring finger, and Ma thumb to little finger. Repetition of the mantra is rhythmic, regulating the breath and soothing the mind. Touching the fingers is tactile and sensory, focusing attention. The meaning of the mantra is "I am truth." For a teenage girl who does not feel confident of her voice or truth but wants to be, this can be a powerful affirmation.

The following resources for working with adolescents with IBS are available:

1. Videos of an 18-year-old female guided in asana and relaxation divided into five parts:
   (a) Video 25.5 Sitting preparation
   (b) Video 25.6 Standing asana practice
   (c) Video 25.7 Kneeling asana practice
   (d) Video 25.8 Supine asana practice
   (e) Video 25.9 Relaxation
2. Figures depicting asana poses useful for IBS
   (a) Figure 25.1 Forward fold sequence
   (b) Figure 25.2 Cat, cow, child sequence
   (c) Figure 25.3 Upward extended arms and legs
   (d) Figure 25.4 Wind-relieving pose
   (e) Figure 25.5 Supine twist
   (f) Figure 25.6 Relaxation pose

Questions

1. Which of the following statements is true?
   (a) The form of the pose is primary; the function of the pose is secondary.
   (b) There is a set sequence of poses that relieves symptoms of IBS.
   (c) All children benefit from vigorous asana practice.
   (d) A body and mind in conflict undermine effectiveness to create desired change.
   (e) Sympathetic activation decreases IBS symptoms of constipation.
2. Which of the following statements is true?
   (a) The sequence order of the practice elements is determined by the intention of the practice and impacts the effect of the practice.
   (b) Starting beginners out with consistent practice elements builds confidence.
   (c) A restless mind and body will likely benefit from more active movement before relaxation.
   (d) Matching the practice to the person's interests will promote adherence.
   (e) All of the above.
3. When referring a client/patient to yoga classes, which of the following are important factors to consider?
   (a) Age of the client
   (b) The training and experience of the teacher
   (c) The temperature of the room during class
   (d) The match between the client's fitness and the vigor of the class
   (e) All of the above

Answers

1. (d): When the body, breath, and mind are focused on the same action/object, they naturally move in a more harmonious direction together. Each element reinforces rather than

Fig. 25.1 Standing forward fold sequence (*tadasana*, *uttanasana*, *ardha uttanasana*). (**a**) Body aligned with weight evenly distributed in tadasana. (**b**) Inhale, sweep arms wide and up with hands to touch, look up. (**c**) Exhale, sweep arms wide and fold, relax head and shoulders, slight bend in knees in uttanasana. (**d**) Inhale, lift halfway back up, hands below knees, neck and spine long in ardha uttanasana. Repeat (**c**) folding down, then (**b**) lifting arms wide and up, and return to standing in (**a**). Repeat sequence 4-6 times

conflicts with the other, and all efforts work toward the same goal rather than in opposition. Yoga practices are intended to create integrated change at the physical, physiological, and mental levels of the person simultaneously.

2. (e): All of the above. Order matters. Preparing and counterposing reduce risk, increase

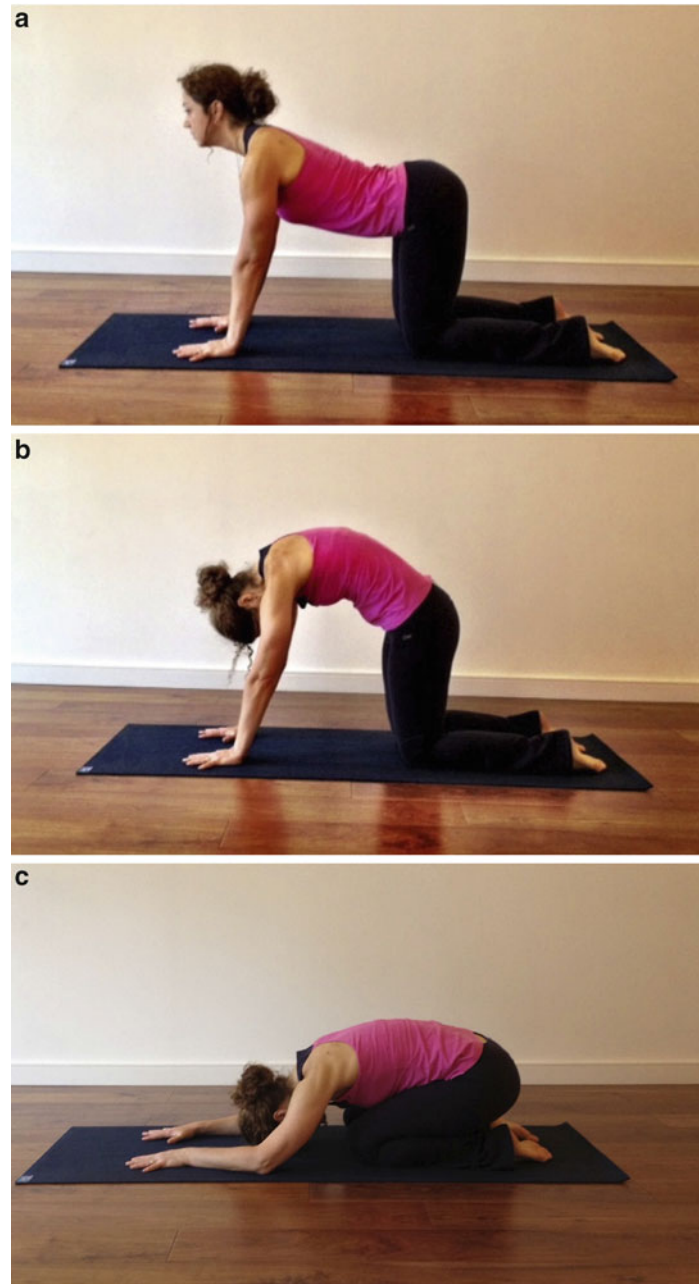

Fig. 25.2 Cat, cow and child pose sequence (cakravakasana). Align the body shoulders over elbows and wrists, fingers spread wide, hips over knees, and feet aligned with knees. (**a**) Inhale, relax belly, lift chest forward, and chin slightly up (cow). (**b**) Exhale, lower head, round spine (cat). (**c**) Exhale, lower head, round spine, lower hips to heels, forehead and forearms to the floor (child). Cakravakasana is a series of poses linked by moving with the breath (**a**) Inhale into cow, (**b**) exhale into cat, and (**c**) exhale into child. Repeat five times. The movement and the breath are slow and rhythmic. The curves of the spine are deepened and reversed. Gentle movement is introduced into the shoulders, elbows, wrists, and hips. The abdomen is gently compressed in child.

benefit, and build balance. Physical and mental restlessness are generally improved by a more physically active practice that demands the mind's attention, uses the big muscles of the body, and requires deeper breathing as this combination naturally increases circulation and clears the stress response hormones (corticotropin-releasing hormone and glucocorticoids) from the system more quickly. Consistency encourages mastery, and people tend to repeat what brings them pleasure.

3. (e): All of the above. It is helpful when referring a client to yoga to be aware that there are different types of yoga classes and that finding

Fig. 25.3 Upward extended arms and legs (urdhva prasarita padasana). (**a**) Exhale, knees into chest. (**b**) Inhale, reach arms above head to the floor and straighten legs toward the ceiling, feet flexed. This is a Counterpose to extend the spine and flatten it toward the floor. To stretch the legs and back. Repeat five times

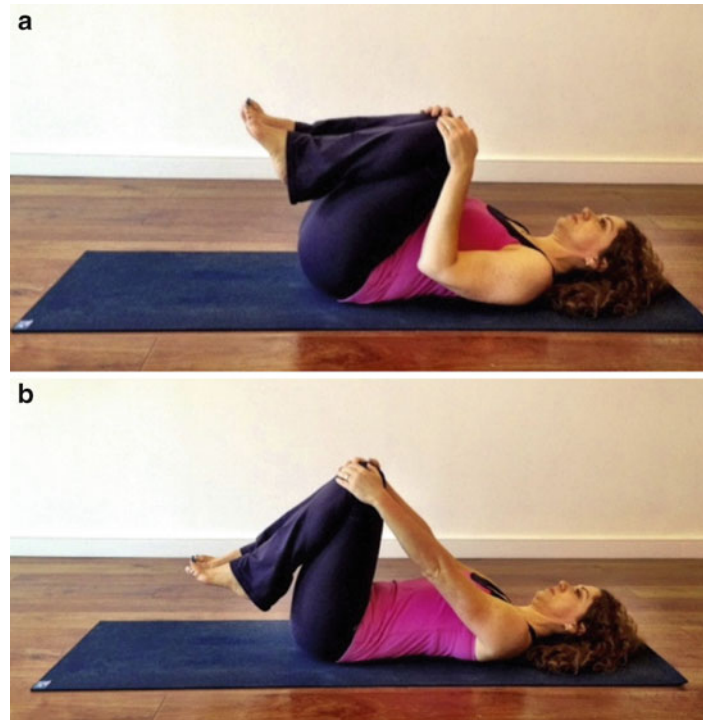

Fig. 25.4 Wind-relieving pose (apanasana). (**a**) Place one hand on each knee; exhale, draw knees into chest, shoulders toward the floor, neck long. (**b**) Inhale, extend arms away, legs and feet stay relaxed. To gently compress the belly while progressively extending exhale and moving rhythmically with the breath. Repeat five times

Fig. 25.5 Supine twist with bent knees (jathara parivrtti). Begin with hands to the side straight out from the shoulders, knees drawn into the chest. Exhale, lower bent knees to the left. Inhale, draw knees back into the chest. Pause one breath. Exhale, lower bent knees to the right. To twist and compress the belly. To gently stretch the hips and low back. Repeat three times each side alternating between sides, then stay on each side for five breaths

Fig. 25.6 Relaxation pose (savasana). Savasana relaxation pose—lie on the floor with arms wide with palms turned up, and feet wide with toes turned out. Use a bolster under the knees to relieve pressure in the lower back and a folded blanket or pillow under the head to support the neck in a neutral position. An eye pillow supports relaxation by turning senses and attention inward. Stay in the pose for 5–10 min. Adopt this pose for guided relaxation and pranayama visualization practices

a good match is important. Encouraging clients to think about how they might benefit from a class and guiding them to seek a class with those qualities can save disappointing and perhaps damaging experiences in classes that are not well adapted to the individual.

Conclusions

Yoga practice is a time-tested method for increasing self-awareness and promoting growth. Such practice fosters strength, flexibility, stability, ease, and balance in the mind and body. The physiological effects of yoga may reduce symptoms of IBS, fibromyalgia, and other functional disorders, as well as alleviate stress, anxiety, and depression. Yoga classes and training programs are widely available with a diversity of class styles to fit the needs of the individual, and specialty classes are available for children of all ages.

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Pharmacotherapy of Functional Disorders in Children

26

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Abstract

Functional disorders (FDs) can often be managed with non-pharmacologic therapies. When drug therapy is employed, selection of medications should be based on the primary underlying cause(s) followed by considerations for symptom abatement. This chapter reviews some of the more common FDs and the evidence for proposed drug therapies. The role of placebo effect is also addressed. A significant focus on gastrointestinal disorders complements the abundance of literature related to this subset of FDs. Consensus guidelines, meta-analyses, and systematic reviews are summarized, and the value of less scientifically rigorous citations is discussed. Drug selection in pediatric FDs is further complicated by added considerations including optimal dose determinations, dosage form challenges, and developmental, pharmacokinetic, and pharmacogenetic variability.

Keywords

Pharmacology • Pharmacotherapy • Psychopharmacology • Antidepressant • Laxative

Introduction

Pharmacology is the study of drug action. More specifically, it is the study of how a drug or substance exerts a biochemical effect on a cell, organ, or other site of action. A related term, pharmacotherapy, is defined as the treatment of disease through the administration of drugs. While functional disorders (FDs) are generally felt to exist in the absence of an organic underlying pathology and are therefore not diseases in the strictest sense, the use of medications to manage FDs straddles aspects of both pharmacology and pharmacotherapy.

A number of core principles related to drug therapy management of FDs should be acknowledged:

- FDs can be treated with non-pharmacologic approaches in many instances, so pharmacologic

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Table 26.1 Functional disorders discussed in this chapter

System	Symptom
Gastrointestinal	Abdominal pain and abdominal pain syndrome
	Abdominal migraine
	Aerophagia
	Cyclic vomiting syndrome
	Retentive fecal incontinence aka encopresis, fecal soiling
	Dyspepsia
	Irritable bowel syndrome
	Adolescent rumination syndrome
	Constipation
Cardiac	Noncardiac chest pain
Musculoskeletal	Fibromyalgia
	Chronic fatigue syndrome
Genitourinary	Vulvodynia and dyspareunia
	Urinary incontinence
	Interstitial cystitis
Respiratory	Vocal cord dysfunction
	Psychogenic sneezing
Miscellaneous	Hirsutism
	Hyperhidrosis

therapy should be reserved for situations in which the non-pharmacologic measures are ineffective or insufficient.

- Given an associated or underlying psychological component with many FDs, therapeutic interventions aimed at managing depression or anxiety should be coincident and undertaken by a clinician properly skilled in managing such conditions.
- Unless otherwise specified, it is assumed that drug therapies employed in the management of FDs are limited to the relief of symptoms and not a resolution of the underlying condition.

There are many FDs described in other chapters throughout this book, some of which will not be discussed here. Throughout this chapter, the authors will identify common FDs for a given body system and then summarize the proposed mechanisms and available evidence supporting drug therapies for a given FD. Table 26.1 lists only those FDs discussed in this chapter. In many cases, supporting information for a given therapy relies on case reports, expert opinion, or personal experience consistent with a lower level of evidence.

Complementary and alternative therapies including herbal products, homeopathic remedies, vitamins, and/or nutritional supplements are widely used by the lay public and on occasion under the direction of medically trained professionals. In addition to a relative paucity of studies of such therapies that employ proper scientific design, most of these substances fall outside the purview of the Food and Drug Administration (FDA) or other regulatory agencies. As such, product consistency and reliable quality are often impossible to validate. Therefore, discussion of complementary/alternative therapies in the management of FDs will be limited. Any extrapolation of the safety and efficacy of different brands or similar products cannot be assured.

Placebo Effect

With the realization that FDs by definition exist in the absence of an underlying physical pathology requiring physical correction, and given the contribution of a psychological component to many FD symptoms, it is reasonable to suspect that a clinical benefit may be derived by some patients by the use of a placebo. In the area of functional gastrointestinal disorders, favorable clinical responses in patients assigned to placebo groups have varied by study and condition but include ranges of 6–72 %, 3–84 %, and 15–72 % with an overall response rate of approximately 40 % in bowel disorder studies [1]. In some studies involving healthy subjects exposed to experimental pain, the magnitude of analgesic response to placebo analgesia—that is, treatment which the patient believes contains an analgesic but is actually placebo—was superior to response to an analgesic medication in patients who enrolled in trials that involved a 1:1 placebo randomization method [2]. These findings support the principle that conditioning (or even a non-pharmacologic approach) can yield a favorable clinical response in some patients as much or more than an actual pharmacologic intervention.

Impact of Psychopharmacology

Investigators have identified an underlying psychological component to many FDs. Therefore, in conjunction with evaluation for possible pathologic causes of a patient's symptomatology, consideration should be given to the potential contribution of a psychological component. This may include, but is not limited to, depressive conditions, anxiety/mood disorders, or other conditions that warrant a systematic evaluation and determination consistent with DSM-V criteria [3].

Selection of the appropriate psychoactive medication for a given FD should involve determination of the underlying psychological condition and consideration for the potential secondary positive or negative efforts of the agent given its mechanism of action [4]. This has particular relevance in patients with depressive conditions for whom a variety of antidepressants may be considered. Such agents may have a pharmacologic basis in mediating serotonin, acetylcholine (i.e., anticholinergic effects), GABA, norepinephrine, and/or other mechanisms. For example, in a patient whose FD involves intestinal spasms, an antidepressant with anticholinergic effects may have a dual benefit. Despite such a pharmacologic basis for selecting a psychoactive agent, evidence-based data validating such a benefit is sparse. For example, a recent Cochrane Review concluded that various psychoactive medications are inconsistently effective in treating gastrointestinal-related FDs [5]. Therefore, optimal use of psychoactive drug therapy in FDs may involve a combination of pharmacologic principles, evidence when available, and/or empiric patient-specific trials. When psychoactive medications are employed, however, it may be difficult to determine if the benefit was derived from improvement of the comorbid psychological condition or from improvement of the functional symptom itself.

Gastrointestinal FDs

Abdominal pain can be caused by a wide variety of functional and pathologic conditions. As with other FDs, evaluation and exclusion of pathologic causes should be undertaken first (Chap. 5). Conditions worthy of assessment include but are not limited to gastroesophageal reflux (GERD), ulcers, Crohn's disease, appendicitis, gallbladder disease, and oncologic conditions. Once pathology is ruled out, symptomatic treatment and/or psychopharmacology, if necessary, is in order. Treatment strategies may include a variety of approaches such as antacids, motility agents, probiotics, and others. The appendix summarizes many citations in the biomedical literature evaluating the pharmacologic approaches to gastrointestinal FDs.

In the 1950s, English pediatrician John Apley first described chronic abdominal pain in childhood and adolescents with no apparent organic causes [6]. In 1988, the International Congress of Gastroenterology met in Rome, Italy, to define criteria to help diagnose irritable bowel syndrome (IBS). The resulting guidance became known as the Rome criteria, the third and current version of which was revised in 2006. The Rome III criteria divided abdominal pain into four distinct categories: function dyspepsia, functional abdominal pain and functional abdominal pain syndrome, IBS, and abdominal migraine (AM) [7]. Summaries of each condition's pharmacologic treatment options are discussed below.

Abdominal Pain and Abdominal Pain Syndrome

Feldman and colleagues evaluated the effects of increased dietary fiber intake in children [8]. This prospective, double-blind, controlled trial randomized 52 pediatric patients to 10 g/day of corn fiber or placebo for a 4-week period. Half the patients in the treatment group (13/26)

reported a 50 % or more decrease in abdominal pain frequency compared to 14 % in the placebo group ($p=0.04$). However, the assessment of pain intensity showed no statistically significant difference between groups. In an uncontrolled trial, Campo and colleagues found that citalopram in doses ranging from 10 to 40 mg daily was associated with a favorable symptomatic response in 84 % of patients by 12 weeks, with 48 % reporting "very much improved" and 39 % being absent of symptoms [9]. Rifaximin was evaluated by Collins et al. in a prospective, randomized, double-blind, placebo-controlled trial of children with varying gastrointestinal FDs including 49 with abdominal pain [10]. Rifaximin 550 mg orally three times daily was given to 23 patients, and 26 patients received placebo. The primary aim of the study was to evaluate lactose breath hydrogen and methane test levels and correlate them to symptom improvement. Symptom improvement was not statistically significant between groups. Teitelbaum and colleagues evaluated the use of amitriptyline or imipramine in 146 patients with a variety of gastrointestinal FDs, including 27 with abdominal pain [11]. Overall response to treatment was favorable in 23/27 patients (85 %). The authors concluded that tricyclic antidepressants (TCAs) may be effective in treating various gastrointestinal FDs in children.

Abdominal Migraine and Cyclic Vomiting Syndrome

AM and cyclic vomiting syndrome (CVS) have been shown to have a strong connection with migraine headaches, and both are considered variants of migraine. However, the term abdominal migraine was developed to assist in recognizing patients characterized with recurrent abdominal pain [12]. The reader is referred to Chap. 5 for detailed reviews of the pathology involved with these conditions.

There are many similarities between CVS and AM despite the fact that they are recognized as two separate entities according to the Rome III diagnostic criteria. However, up to 80 % of CVS patients have abdominal pain, and up to 72 % of AM patients have vomiting episodes making a differentiation difficult [12]. There has been a question of whether they are actually distinct diagnoses [12]. Taking this into consideration when looking at available literature for treatment of CVS and AM, delineation between the two conditions is not clear. Many authors have used the terms interchangeably with respect to treatment. Therefore, treatments for CVS and AM are similar and often parallel. Pizotifen and cisapride have been utilized with success in reducing acute events of abdominal migraine and CVS. But as these drugs are not available in the USA, they will not be reviewed but referenced [13–18].

Cyclic Vomiting Syndrome

Treatment is typically considered abortive/supportive or prophylactic with literature support in the form of retrospective reviews, case reports, and series and limited controlled trials.

Abortive/Supportive

Vanderhoof and colleagues conducted two studies evaluating erythromycin through a prospective case series. Therapy was found to be effective (≥50 % improvement) in 62 % and 67 % patients according to initial response [19, 20]. Promethazine, ondansetron, and oxazepam were shown to be effective for relieving episodes, while a multimodal treatment approach alleviated intensity and frequency in a single case report [21]. Phenobarbital was noted to cause improvement of symptoms in 100 % of patients ($n=14$) with 11/14 showing complete resolution of symptoms and 3/14 showing marked improvement in a study by Gokhale and colleagues [22]. Khasawinah et al. found dexmedetomidine to be effective in 4 of 5 episodes in three patients with CVS [23]. Sumatriptan has also been used for abortive therapy in CVS in a 14 year old girl with complete improvement soon after administration [24]. A prospective, non-blinded, non-placebo study also reported patient response (≥50 % improvement) 75 % of the time (9/12 patients) and an overall attack response of

54 % (19/35 attacks) with sumatriptan [25]. Lastly, clonidine has been employed by several investigators. Abraham and colleagues showed complete resolution of 6/6 episodes in a single patient using clonidine [26], and Palmer et al. reported improvement in CVS following the use of intravenous midazolam and clonidine [27].

Prophylaxis

More literature exists for use of prophylactic treatment in CVS but is far from plentiful. Among the most described therapies are amitriptyline [17, 28–34], cyproheptadine [16, 28, 33], and propranolol [16, 17, 33, 34] with all having at least a modest response to complete resolution of symptoms. Less well-studied medications include topiramate [32, 35], coenzyme q-10 [31, 32], L-carnitine [32, 36], valproate/phenobarbital [37, 38], and nebivolol [30] showing some benefit in patients. Refer to the appendix for more detailed information for each medication.

Medication Combinations

Various studies have used a combination approach for acute treatment and prophylaxis. Due to the heterogeneity of the respective citations and the variable responses seen with unspecified combinations, the reader is directed to the appendix to consider the relative merit of each citation and the medications employed.

Guidelines

The North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition (NASPGHAN) released guidelines in 2008 on the diagnosis and management of CVS [39]. Recommendations for management are based on lower quality retrospective reviews or case reports or poor quality controlled studies (evidence Level II-1 to Level II-3, Grade D). A greater degree of evidence is available to support prophylactic therapies. In patients 5 years of age or younger, cyproheptadine is recommended as the first choice in prophylaxis. When children are older than 5 years of age, amitriptyline is the treatment of choice. Propranolol is the second line for both age groups. For acute management, the guidelines recommend a trial of a $5HT_{1B/1D}$ agonist, a "triptan," in patients 12 years of age or older to attempt aborting the attack. Other supportive (not considered abortive) therapies can be utilized such as $5HT_3$ antagonists, sedatives, and analgesics.

Abdominal Migraine

A retrospective review of patients with abdominal migraine published by Worawattanakul and colleagues reviewed use of propranolol and cyproheptadine [40]. Eighty-three percent of patients receiving propranolol had at least a fair response to treatment compared to 83 % of patients in the cyproheptadine group. Both groups had a decrease in the number of attacks. There was no statistical significance noted between groups. Intravenous valproic acid was also reported to have benefit in two patients with refractory abdominal migraine by resolving symptoms [41]. However, one patient had to discontinue therapy due to adverse effects. As previously noted, such a small case report has limited generalizability unless larger studies validate similar findings.

Evidence is severely limited for the management of AM. As stated previously, many studies have used the terms CVS and AM interchangeably, and therefore, many of the treatment approaches have been used in both syndromes. Anecdotal evidence exists for using treatments used in migraine for treating AM [42].

Aerophagia

In separate trials by Chitkara, Hwang, and Loening-Baucke, these investigators have conducted studies involving education, behavioral therapy, and the use of omeprazole, aluminum hydroxide, simethicone, chlordiazepoxide, metoclopramide, dicyclomine, clonazepam, milk of magnesia, and polyethylene glycol [43–45]. Each trial had one or more limitations including small sample size (45, 22, and 5 subjects, respectively), retrospective review or observational study, heterogeneous outcome measures, and limited

statistical application. Chitkara and colleagues reported that of 45 patients retrospectively evaluated for aerophagia, 12 had a documented follow-up visit [43]. Of those follow-up patients, four (one treated with a combination of medications, three who received education alone) reported improvement. In 2011, a consensus group led by Benniga advised and Bredernoord affirmed that although anti-gas agents such as simethicone have not been rigorously studied in patients with aerophagia, these medications should be considered first-line treatment based on their known effects of reducing surface tension and gas formation in the intestine [46, 47].

Encopresis

Pharmacotherapy (e.g., with laxatives) of encopresis without constipation, also known as functional nonretentive incontinence, has not been shown to add benefit to behavioral therapy [48–50].

Treatment of constipation and encopresis is an involved process usually requiring education, disimpaction, maintenance therapy (preventing the reaccumulation of stool), and behavioral treatment [51]. Brazzelli et al., in a Cochrane Review, report improvement of incontinence when behavior modification is combined with laxatives over use of laxatives alone [52]. VanGinkel and coworkers prospectively evaluated the impact of adding lactulose to a biofeedback regimen in a randomized, non-blinded study of 48 children [53]. Both groups had similar statistically significant reductions in episodes compared to baseline. However, there were no differences between groups with regard to colonic transit time or other measures including anorectal manometry. In 2002, Borowitz et al. conducted a prospective study of 87 children with encopresis [54]. Patients were randomized to one of three groups: intensive medical therapy (IMT) involving disimpaction with enemas, laxatives (e.g., milk of magnesia and senna) titrated to effect, and rescue enema or suppository if no bowel movement in 48 h; IMT plus behavior management including enhanced toilet training; and IMT plus behavior management plus biofeedback. Significant improvements in the primary outcomes of daily soiling frequency, number of bowel movements in toilet per day, and total cure (defined as absence of soiling in a 2-week period) were noted in all groups, but there were no statistically significant differences between groups. In both studies, the addition of pharmacologic therapies to behavioral interventions yielded no statistically significant incremental benefit.

Dyspepsia

Pharmacotherapy of dyspepsia not associated with GERD or other pathology in children has yielded results similar to other FDs, namely, the absence of a consistent response from an array of therapeutic modalities. A histamine-2 receptor antagonist, famotidine, was compared to placebo in a prospective, blinded, placebo-controlled crossover study [55]. In these 25 patients, there was no statistically significant change in a quantitative abdominal pain score between groups, but more patients (68 vs. 12 %) reported improvement with famotidine over placebo according to a subjective assessment. Studies involving TCAs have had mixed results. In 2009, Saps and colleagues compared amitriptyline to placebo in a group of 43 patients with IBS, functional abdominal pain, or functional dyspepsia [56]. More patients in the amitriptyline group self-reported feeling better compared to placebo (63 vs. 57.5 %), but this finding did not reach statistical significance. As noted previously, Teitelbaum and Arora published their retrospective review of 146 patients who received either amitriptyline or imipramine for IBS, dyspepsia, or functional disorder of the intestine [11]. For the subset of patients with functional dyspepsia, 12 of 16 patients who received either TCA were considered to have responded well. Horvath and colleagues performed a meta-analysis on the use of lactobacillus in a variety of FDs, including functional dyspepsia [57]. In the three outcomes related to frequency, intensity, and overall abdominal pain, there were no statistically

significant differences between treatment with lactobacillus and placebo. In the Collins et al. study of rifaximin vs. placebo, 11 patients with dyspepsia received treatment [10]. No statistically significant symptom outcomes favoring treatment were identified in this study.

The role of montelukast in dyspepsia was evaluated in a randomized, double-blind, placebo-controlled crossover trial in 40 pediatric patients with dyspepsia and duodenal eosinophilia [58]. Patients received either montelukast 10 mg or placebo orally daily for 14 days. After day 14 assessment, patients were converted to the alternate group and continued for another 14 days. Ranitidine was co-administered throughout the study to all but two patients. Evaluation of symptomatic response to therapy included a 5-point Likert scale; a favorable response was defined as grades 3–5. In the first 14 days, montelukast-treated patients reported 62.1 % response compared to 32.4 % response in the placebo group ($p<0.02$). Mixed response without statistical significance was noted after the crossover period ended. In a subgroup analysis of patients with marked eosinophilia, symptomatic response favored montelukast-treated patients (68 vs. 32 %, $p=0.01$). Cisapride and tegaserod have also been employed in pediatric patients with functional dyspepsia [59, 60]. However, both products are no longer available in the USA due to adverse effects including QTc prolongation with cisapride and cardiovascular events with tegaserod.

Irritable Bowel Syndrome

Children and adults with IBS have evidence of visceral hypersensitivity, resulting in pain [61]. This may stem from a dysregulation of the communication between nerves in the abdominal area and the central nervous system. The cause of visceral hypersensitivity may be due to numerous factors, including physiologic stimuli, noxious stimuli, altered motility, and/or psychological stress. Treatments aimed at mitigating the visceral hypersensitivity and bacterial overgrowth have been devised.

The placebo response rate in treating IBS has been reported to be as high as 37.5 % in one meta-analysis [62]. As discussed in previous sections, TCAs including amitriptyline and imipramine have had mixed results in pediatric studies involving more than one gastrointestinal FD [11, 56]. One study by Bahar et al. specifically evaluated the use of amitriptyline in adolescents with IBS [63]. The study was well designed as a prospective, double-blind, randomized controlled trial using a weight-based dose of amitriptyline or placebo for 8 weeks. Primary outcomes were quality of life score improvement and reduction in IBS-associated diarrhea. Both outcomes showed statistically significant changes favoring the amitriptyline-treated patients at various times during the study. In addition to TCAs, selective serotonin receptor inhibitors (SSRIs), monoamine oxidase inhibitors (MAOIs), and mirtazapine have been shown to improve IBS symptoms in adults, but supporting data in pediatric patients is lacking [64–67]. Lubiprostone is FDA approved in adults for constipation-related IBS and idiopathic constipation, but no supporting pediatric data exists [68].

Management of irritable bowel syndrome with diarrhea as a predominant symptom (IBS-D) has included several additional therapies. Serotonin antagonists including ondansetron and alosetron have been studied in adults, but pediatric data is lacking, and alosetron has limited availability due to severe ischemic colitis and other complications associated with severe constipation [68, 69]. Other therapies including antidiarrheals such as loperamide and antispasmodics may have utility in IBS-D. However, such agents do not have a sufficient evidence base to recommend use in pediatric patients at this time [68, 69].

Probiotic efficacy appears to be species specific. The benefit of *Lactobacillus rhamnosus GG* treatment is supported by a meta-analysis and two placebo-controlled trials evaluating pediatric functional abdominal pain that included patients with IBS [57, 70, 71]. Compared to placebo, outcomes related to pain intensity, pain frequency, and overall response favored lactobacillus treatment. Martens and colleagues evaluated a German

Escherichia coli-based product (Symbioflor 2) in pediatric patients with IBS [72]. The investigators noted statistically significant improvements in pain and stool frequency. Presently, this product is not distributed in the USA. Probiotics other than *lactobacillus GG* have shown efficacy in adults with IBS [73]. Kline and colleagues evaluated the use of peppermint oil capsule in 42 children with IBS [74]. Patients were randomized in a double-blind fashion to receive placebo or a fixed weight dose (187 or 374 mg) three times daily for 2 weeks. Outcomes of change in symptom severity scale and, specifically, pain symptoms favoring peppermint oil were statistically significant. As previously described, Collins et al. evaluated rifaximin in several pediatric GI FDs, of which 41 IBS patients were included (with 26 patients in the placebo group) [10]. The outcome of symptom improvement was found to be not statistically different between treatment and placebo groups. Pantoprazole has also demonstrated effectiveness in adult patients with overlapping symptoms of gastroesophageal reflux disease and IBS [75]. Cisapride and tegaserod have been employed in pediatric patients with IBS [60, 76–79]. However, since neither agent is presently commercially available in the USA, they will not be further discussed.

Adolescent Rumination Syndrome

The effortless regurgitation of food into the mouth is a common characteristic of rumination syndrome. It affects infants and the developmentally disabled more often than normal children, adolescents, and adults. It has been postulated that abnormal relaxation of the lower esophageal sphincter, a classic sign of GERD, also plays a role in rumination. Chial et al. retrospectively reviewed the records for 147 patients between 5 and 20 years old who were diagnosed with rumination syndrome [80]. While these authors reported various demographic, diagnostic, and treatment parameters, they did not conduct an interventional study nor did they make conclusions regarding treatment. However, it was mentioned that behavioral treatment including habit reversal is their primary intervention and that any comorbid medical or psychological conditions would require additional intervention. Khan and colleagues reported that a combination of medications, nutritional support, biofeedback, relaxation techniques, and cognitive therapy resulted in improvement in 10/12 patients [81]. Six patients were reported to have received anxiolytic or antidepressant medications, but the specific agents or regimens were not described. The authors found that multimodal therapy was effective. However, conclusive findings with regard to medication selection, dose, or effectiveness cannot be made from the information provided. Tack et al. cited information from an unpublished abstract describing the use of baclofen in pediatric patients with rumination [82]. The abstract authors reported a 50 % decrease in regurgitation symptoms and a 65 % reduction in rumination episodes. No statistical analysis was employed. Levosulpiride is a selective antagonist of the dopamine D2 receptor with activity on both central and peripheral nerves and possessing neuroleptic and prokinetic effects. It has been shown to be effective when combined with psychotherapy in adult patients [83]. However, it has not been evaluated in pediatric patients and it is not yet commercially available in the USA.

Constipation

When the defecation pattern of children deviates from what is considered to be a normal pattern, parents and caregivers may express concern and alarm and/or request medical intervention. Beyond the neonatal period, the most common cause of constipation is functional [84]. Treatment of functional constipation involves a stepwise approach including exclusion or resolution of impaction followed by maintenance therapy, education, and follow-up. Treatment options for constipation include a wide range of agents, doses, and routes of administration. The NASPGHAN has issued consensus guidelines for the treatment of functional constipation [84]. Stimulants, osmotic agents, and lubricants constitute the three major categories of therapy. Specific doses,

side effects, and treatment considerations are well described within this document, and the reader is directed to this citation for additional information. Patient preferences or patient-specific factors should guide the clinician's choice in therapy given that equal benefit should be derived from an appropriate dosed and administered product regardless of type or route of administration.

Cardiac FDs

Noncardiac Chest Pain

NCCP is rarely seen in children and is considered by many to be a functional gastrointestinal disorder given the propensity for patients to have coinciding GI complaints and a negative cardiac history by exam and/or angiography (Chap. 3). Such patients may also have been evaluated for conditions with overlapping symptoms such as GERD. Mayou and colleagues identified more than half of their adult cardiac clinic patients who were evaluated for chest pain as having no ischemic heart disease or other physical cause [85]. In a latter study, the same author and his collaborators evaluated 133 adult patients with NCCP (69 via angiography, 64 via clinical assessment) [86]. A subset of patients who had persistent NCCP following reassurance by a cardiologist participated in a trial of cognitive behavioral therapy (CBT). Fourteen of 16 NCCP patients reported substantial improvement of symptoms following CBT compared to no improvement in 17 control subjects. Similarly, Mudipalli and colleagues evaluated 69 adult patients with functional chest pain [87]. Eighty-two percent of patients had an underlying gastrointestinal FD including IBS. These findings support the suggestion that identification of another primary cause of NCCP, especially consideration of psychological conditions, is of paramount importance. Pharmacologic treatments should, therefore, be based on the underlying diagnosis. Refer to discussions of gastrointestinal FDs and psychopharmacology.

Musculoskeletal FDs

Fibromyalgia

FM is considered to be part of a spectrum of conditions that also includes chronic fatigue syndrome (CFS) and depression [88] (Chap. 11). FM patients are also noted to have overlapping FDs including IBS [89]. Etiology of FM is multifactorial and may involve psychological stressors, sexual abuse, learning difficulties, and/or genetic predisposition. Treatment involves a range of interventions including education, aerobic exercise, behavioral therapy, and complementary/alternative medicine. Pharmacologic therapy in adults has included gabapentin, pregabalin, and duloxetine. However, there are no well-designed studies of drug therapy in children with FM. Mariutto and colleagues reported their experience with fluoxetine in an open-label trial in ten female adolescents with juvenile FM [90]. Only four patients were compliant with the 12-week protocol, but those patients demonstrated improvements in pain score and other outcome measures. However, the quality of their study precludes interpretation and broader applicability. The effectiveness of analgesics and NSAIDS in improving FM symptoms has not been established, and only limited data exists to suggest the possible benefit of cyclobenzaprine or amitriptyline [88].

Chronic Fatigue Syndrome

As previously noted, CFS is one of the spectrum of coexisting or similar FDs (Chap. 8). The etiology of CFS has not been precisely determined, and diagnosis is contingent on the exclusion of other causes such as psychiatric diagnoses, bacterial infection, Epstein-Barr virus infection, or other immunologic conditions [91]. A number of drug therapies have been tested over the years; some aimed at treating a potential etiology without success, including various immunotherapies, corticosteroids, and antidepressants.

The reader is referred to a summary of previous therapeutic interventions summarized by Afari [91]. More recently, Fagermoen and colleagues evaluated the utility of oral clonidine in a pilot study involving five adolescents with CFS [92]. The authors concluded that clonidine may have benefit in reducing the patients' orthostatic intolerance with minimal side effects. Further study is required to validate these preliminary findings. Emphasis on addressing underlying treatable conditions and reducing painful symptoms, if present, appears to be the best current recommendation for pharmacologic management of CFS.

Vulvodynia and Dyspareunia

While predominantly seen in adults, this condition may in theory affect sexually active teens. Proposed etiologies of localized provoked vulvodynia (LPV) include interstitial cystitis, hypertonia, localized mast cell dysregulation or neuroproliferation, and underlying psychological conditions. In 2011, Khandker and colleagues found an increased incidence of mood or anxiety disorder in women with vulvodynia compared to a general population control group (26.7 vs. 11.3 %, respectively) [93]. Surgical intervention and physical therapy have been employed. Pharmacologic treatments with antidepressants, anticoagulation with enoxaparin, local anesthetics, and direct muscle relaxants (botulinum toxin) have yielded variable success. Farajun and colleagues evaluated 40 women with vulvodynia who received either enoxaparin 40 mg subcutaneously or saline placebo daily for 90 days [94]. Statistically significant improvements in sensitivity and pain reduction were noted in the treatment group as was a histological improvement of biopsy findings. The proposed mechanism for enoxaparin's effect was not systemic antithrombosis but a local reduction in heparanase activity, thus preventing degradation of vestibular stroma and increased nerve fiber penetration in the vulvar epithelium. Pelletier et al. evaluated the local use of botulinum toxin A (BTA) in 20 women with refractory LPV [95]. The patients received 100 units of BTA and were evaluated at 3 and 6 months after treatment. Eighty percent of patients reported improvements in pain at 3 and 6 months postinjection; 13 of 18 patients were able to resume sexual intercourse following treatment. Foster and colleagues concluded that oral desipramine and topical lidocaine alone or in combination were ineffective in reducing vulvodynia pain in a placebo-controlled 12-week trial [96]. Table 26.2 summarizes the citations reviewed herein.

Table 26.2 Summary of vulvodynia/dyspareunia citations

Intervention	Data	Citations
Enoxaparin	CT	Farajun et al. [94]
Botulinum toxin A	CS	Pelletier et al. [95]
Desipramine/lidocaine	CT[a]	Foster et al. [96]

CR case report, *CS* case series, *CT* controlled trial
[a]Neutral/negative findings

Interstitial Cystitis

The causes of interstitial cystitis (IC) may be pathologic, related to sexual trauma, or idiopathic. While there are a number of reviews and case reports describing treatment of IC, most of these citations involve adults. Reviews by Mattox and Yoost et al. which summarized IC treatment in children and adolescents refer to a small number of pharmacologic modalities that have been employed with or following other multimodal approaches [97, 98]. Drug therapies employed have included pentosan polysulfate, hydroxyzine, cimetidine, cyclosporine, amitriptyline, and nortriptyline. Intravesicular treatments containing heparin, lidocaine, and dimethyl sulfoxide have also been employed with success. Other intravesicular therapies have been used but with little clinical benefit [97, 98].

Urinary Incontinence

Two main types of functional urinary incontinence (FUI) have been characterized: bladder overactivity and dysfunctional voiding [99] (Chap. 6). Etiologic factors and treatment options for these

two groups differ. Treatment of FUI is multifaceted. Since constipation is a common finding in such children, initial management with laxatives and an ongoing bowel program (with training and/or medications) is important [100]. The reader is referred to the discussion within this chapter and secondary sources for guidance on pharmacologic management of constipation. Other interventions may include cognitive and behavioral therapy, education, biofeedback, and relaxation techniques. In patients with bladder overactivity, the use of antimuscarinic agents may reduce involuntary detrusor contractions. Oxybutinin has long been the drug of choice for this condition [99]. However, systemic anticholinergic side effects including xerostomia and constipation may limit its utility in some patients. Tolterodine also has antimuscarinic properties and appears to have less effect on salivary activity. It has been employed in adults with overactive bladder, and its use in children has been evaluated in several small studies. Bolduc and colleagues evaluated tolterodine use in 34 children who received multimodal interventions and did not tolerate oxybutynin therapy [101]. Patients aged 5–9 years received tolterodine 1 mg orally twice daily; children 10 years and older received 2 mg twice daily (a common adult dose). Side effects reported during tolterodine therapy were less than those previously reported while patients received oxybutynin. Statistically significant reductions in xerostomia were noted; other findings did not reach statistical significance due to the lower frequency and small population.

Management of dysfunctional voiding, in which underactive or uncoordinated detrusor activity exists, may in contrast theoretically involve cholinergic agents. However, the use of such medications including bethanechol has not demonstrated effectiveness [100]. Alpha-adrenergic blockers such as doxazosin are commonly used in adults, such as males with benign prostatic hypertrophy and other functional bladder disorders. Data supporting use of these agents in children is limited, however. Kramer and colleagues employed doxazosin 0.5 mg daily in a randomized, double-blind, placebo-controlled trial in 38 children with voiding dysfunction [102]. The primary outcomes related to incontinence suggested improvement in doxazosin-treated patients but were not statistically significant. Similarly, Austin and colleagues observed improvements in bladder function in 17 children treated with doxazosin for a variety of urologic conditions including dysfunctional voiding [103]. Bladder symptoms or function improved in 82 % of patients, but the absence of a control group and randomization precluded statistical analysis. BTA injection has been employed by several investigators for management of refractory cases of lower urinary tract dysfunction, but limitations of study quality and size preclude conclusive recommendations for this therapy presently [100].

Respiratory FDs

Vocal Cord Dysfunction

Presenting symptoms of vocal cord dysfunction (VCD) may be confused with uncontrolled asthma (Chap. 4). A number of disorders including major depression, anxiety, obsessive compulsive disorder, other psychiatric conditions, and stress temporally associated with competitive sports have coincided with VCD [104]. The symptoms of VCD are often brief and self-resolving. Asthma-related drug therapies are helpful if the patient has underlying asthma. Anxiolytics and/or other psychoactive drugs may address an underlying psychiatric component. Novel therapeutic approaches have included topical lidocaine and local botulinum toxin injections. BTA prevents acetylcholine release at presynaptic nerve terminals, which effectively results in a denervation. Its utility has also been shown in the treatment of blepharospasm, skeletal muscle contractions, dysphonia, and for cosmetic purposes. Maillard and colleagues described a severe case of VCD in an adult woman which required intubation [105]. Following 6 days of mechanical ventilation and the failure of other therapies, intralaryngeal injection of BTA was performed under general anesthesia. The patient experienced slow but

Table 26.3 Summary of vocal cord dysfunction citations

Intervention	Data	Citations
Botulinum toxin A	CR CS	Maillard [105] Marion [109]
Lidocaine	EO	Kenn [110]
Heliox	EO	Reisner [111]
Amitriptyline	CS	Varney [106]
Ipratropium	EO	Weinberger [107]

CR case report, *CS* case series, *EO* expert opinion, *CT* controlled trial

progressive improvement thereafter and was discharged 5 days later. A trial of low-dose amitriptyline (mean 20 mg/day) coincident with speech therapy in 62 adult patients indicated clinical response in VCD symptoms in 82 % and 94 % of female and male subjects, respectively [106]. The authors postulated that the anticholinergic and/or muscle relaxant effects of amitriptyline may have been a contributor to response. Weinberger and Abu-Hasan reported anecdotal favorable experience with ipratropium in patients with exercise-induced VCD, but this has not been validated in controlled trials evaluating functional VCD [107]. In the absence of underlying reactive airway disease, bronchodilator therapy such as albuterol has not benefitted patients with VCD [108]. Other therapies which address comorbid conditions may help to improve VCD, but the benefit is related to the underlying condition, not VCD itself. Table 26.3 summarizes the citations reviewed herein.

Psychogenic Sneezing

First described in 1949, psychogenic sneezing (PS) differs from the usual protective response associated with irritant triggers in the nasal passages (Chap. 8). Primary treatment is widely accepted to involve psychotherapy as an underlying psychological component is involved [112]. Several case reports describe pharmacotherapeutic interventions. Sulemanji and colleagues reported their evaluation of an 11-year-old female with PS [112]. During diagnostic procedures, administration of a local anesthetic appeared to later reduce the frequency of sneezing. The authors suggested that consideration of local anesthetic administration as part of a PS workup may be reasonable. Other investigators have employed and suggested the use of intranasal local anesthetics as a tool in the differential diagnosis process, but no evidence yet validates the effectiveness of such therapy in treating PS. Guner and colleagues reported resolution of PS in a 12-year-old girl who was already receiving sertraline 50 mg/day for a previously diagnosed anxiety disorder [113]. Haloperidol 1 mg daily was initiated and the patient's signs were reduced in 3–4 days with complete resolution in 2 weeks. Haloperidol was discontinued after 1 month, and the patient remained PS symptom-free with ongoing sertraline and psychotherapy at 6-month follow-up.

Miscellaneous FDs

Hirsutism

The presence of excessive hair in areas of the female body affected by androgenic hormones is usually attributable to pathologic conditions such as hyperandrogenism, polycystic ovary syndrome (PCOS), or congenital adrenal hyperplasia [114]. These conditions are beyond the scope of this discussion. However, absent other pathologies, idiopathic hirsutism qualifies as a functional disorder. It is usually benign and largely cosmetic but treatment usually involves hormone therapy. Oral contraceptives are generally the drug of choice, and progestin-containing products with neutral or antiandrogenic properties are preferred [115]. Antiandrogenic agents are generally reserved for patients with a pathologic androgen imbalance and are therefore not reviewed in this FD focused discussion. The reader is directed to evaluation and treatment guidelines for hirsutism in premenopausal women developed by The Endocrine Society [115].

Hyperhidrosis

Excessive sweating disproportionate to the need for thermal regulation not associated with other pathologies is considered primary hyperhidrosis

and mostly involves the axillae, palms, soles, and face. The negative impact of hyperhidrosis is mostly emotional and social. Usual onset of symptoms occurs between ages 14 and 25 years [116]. Various topical and systemic therapies have been evaluated. Topical astringents with aluminum salts are logical first-line therapies as these form the basis for most commercial antiperspirants. In 2008, Gelbard and colleagues reviewed pharmacologic treatment options, and the reader is directed to this article for specific details on agents, doses, and evidence [116]. The range of topical agents employed include aluminum chloride in various alcohol, salicylic acid, and water-based dilutions with or without occlusive dressing. More potent agents including formaldehyde have been effective but associated with unfavorable local side effects. Topical application of anticholinergics such as glycopyrrolate has been described in several adult case reports. BTA injection and iontophoresis with or without anticholinergic complete the list of topical intervention options. Systemic anticholinergics including glycopyrrolate and propantheline may be effective but have been limited by dose-related adverse effects. Oxybutinin appeared to have a serendipitous benefit resolving hyperhidrosis in a woman with urge incontinence. Other systemic medications including diltiazem, clonidine, alpha-adrenergic blockers, and benzodiazepines have been employed but supporting literature does not include pediatric patients.

Questions

1. Which of the following statements regarding pharmacotherapy for FDs is most appropriate?
   (a) Pharmacotherapy should always be first line for symptomatic control.
   (b) Pharmacotherapy should be used in conjunction with other treatment modalities such as behavioral modification.
   (c) In the absence of organic disease, drug therapy has no role in FDs.
   (d) Once started, pharmacotherapy should continue indefinitely.
   (e) All of the choices are inappropriate.
2. Selection of psychoactive medications for certain FDs is most strongly supported by which principle?
   (a) In some FDs, the side effect profile (e.g., anticholinergic properties of amitriptyline) of a psychoactive medication may have the added benefit of offsetting certain symptoms.
   (b) Anxiolytics should be considered for any FD.
   (c) Psychoactive medications should only be considered if symptom-based drug therapy previously failed.
   (d) Only a psychiatrist should prescribe psychoactive medications following a full diagnostic workup.
   (e) None of these principles are valid.
3. In the absence of scientifically rigorous clinical data supporting the use of a medication in a pediatric patient, which of the following principles best apply?
   (a) Evidence supporting a given drug therapy in adults always justifies use of the drug in children.
   (b) The dose of medication that would be safe and effective for a child is the fraction of "weight divided by 150 lb."
   (c) Constipation in a child is best treated with rectally administered medications to optimize local effects.
   (d) Best clinical judgment accounting for risk vs. benefit of an unproven therapy in a pediatric patient should be a primary consideration prior to initiating therapy.
   (e) In the absence of scientifically rigorous clinical data, drug therapy should not be administered for the treatment of a child with an FD.

Answers

1. (b): As functional symptoms often improve or resolve with the use of non-pharmacologic interventions, pharmacotherapy should be offered following or in conjunction with non-pharmacologic interventions.

2. (a): When non-pharmacologic interventions are insufficient, psychoactive medications can be prescribed by appropriately trained primary care providers for patients whose FDs are triggered or perpetuated by anxiety or depression. As suggested by the scenario within this question, it may be helpful to consider the side effect profile or the specific mechanism of action (i.e., secondary neurotransmitter effects) that may contribute to symptomatic improvement. Amitriptyline has a high level of anticholinergic effects compared to some other TCAs and thus may have added benefit in treating patients with IBS-D and coincident depression.
3. (d): Given the lack of high quality evidence of treating FDs in children, pharmacotherapy should be initiated with careful consideration. Altered pharmacokinetics and pharmacodynamics may result in a different response to drug therapy in children, so one should not extrapolate the safety for a given drug in children despite evidence of safety in adults. Dosing of medications in children and adolescents is often NOT a direct proportion of age, weight, or other anthropometric measure. Certain routes of administration (i.e., rectal) may be effective but far less acceptable/favorable for use in children. When all available factors and information have been considered, a best judgment determination of drug selection and dosing in a child may be necessary.

Summary and Conclusion

Functional disorders in children involve a constellation of symptoms and body systems, often with significant overlap. Psychological disorders are frequently identified as comorbid or primary factors. Exclusion of a range of pathologic conditions is a key element in managing FDs. With regard to treatment of FDs, pharmacotherapy should be considered only after sufficient trials of non-pharmacologic interventions have demonstrated failure or incomplete results. When employed, pharmacotherapy should be designed to address the primary problem(s) first followed by targeted symptom management. Numerous examples demonstrate that pharmacologic principles and treatment approaches addressing proposed underlying mechanisms of FD symptoms are not always successful. In other circumstances, the selection of psychoactive medications based on coincident secondary effects (e.g., anticholinergic side effects from amitriptyline) has not reliably demonstrated the expected effect.

The guiding principles of drug therapy in children must include all of those that apply to adults AND considerations of altered pharmacokinetics, unique dosing and administration considerations, and safety/toxicity risks. When scientific evidence is available to address the selection and dosing of medications in treating pediatric FDs, the caregiver must always consider the context, quality, and situational applicability of the information to a given patient care situation. When supporting literature is not available and empiric pharmacotherapy is chosen, consideration must be given to cautiously select the drug with the widest margin of safety, a reasonable pharmacologic basis for use, and the lowest possible effective starting dose. Clinicians must account for dosing factors based on age, weight, drug interaction potential, drug allergy history, organ function, and drug–disease interactions. The acknowledgment that "children are not just small adults" is no less important in treating functional disorders than in organic disorders.

Studies Evaluating Medications in Recurrent Abdominal Pain

Medication	Intervention	Control intervention	Outcome measured	Results	Efficacy
Dietary fiber [8]	Prospective study using 10 g/day corn fiber × 4 weeks after 2 week run-in period ($n=26$) in patients with at least one episode of unexplained abdominal pain per week × ≥2 months	Placebo ($n=26$)	**Decrease in attacks**	**Decrease in attacks**	
			Frequency	**Frequency**	$p=0.04$
				Fiber 13/26	More effective
				Placebo 7/26	
			Intensity	**Intensity**	
				Not reported	Not reported
Citalopram [9]	Prospective open-label, flexible dose study in patients with functional recurrent abdominal pain × 12 weeks (range: 10–40 mg) ($n=25$)	None	**Clinical Global Impression Scale-Improvement** Self- and parent-reported abdominal pain, anxiety, depression, other somatic symptoms, and functional impairment	**Clinical Global Impression Scale-Improvement** Week 8: 80 % response Week 12: 84 % response Very much improved: 48 % Absent symptoms: 39 %	N/A
Rifaximin [10]	Prospective, double-blind, placebo-controlled study of patients with functional dyspepsia ($n=11$), IBS ($n=41$), functional **abdominal pain** ($n=23$), or abdominal migraine ($n=0$) that were given rifaximin ($n=49$) Rifaximin: 550 mg orally three times daily × 10 days	Placebo ($n=26$)	**Symptom improvement (Overall and individual symptoms)** **Hydrogen levels** (not reported here) **Methane levels** (not reported here)	**Symptom improvement** Specifics not mentioned in study	All values not significant between groups
Tricyclic antidepressant (TCA) [11]	Retrospective review of patients prescribed with amitriptyline or imipramine ($n=146$) for irritable bowel syndrome, dyspepsia, **functional disorder of the intestine**	None	**Overall response to treatment**	**Overall response to treatment** **Responded**: 78.6 % for average of 10.73 months *Amitriptyline* Responded well: $n=40$ (FAP $n=9$) Did not respond: $n=8$ (FAP $n=1$) Stopped due to adverse effects: $n=3$ (FAP $n=0$) *Imipramine* Responded well: $n=41$ (FAP $n=14$) Did not respond: $n=5$ (FAP $n=2$) Stopped due to adverse effects: $n=2$ (FAP $n=1$)	TCAs effective in treating function gastrointestinal disorders in pediatric patients (general)

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Medication	Intervention	Control intervention	Outcome measured	Results	Efficacy
Cyproheptadine [117]	Prospective, randomized, double-blind, placebo-controlled trial of patients with functional abdominal pain that were given cyproheptadine ($n=15$) for 2 weeks Cyproheptadine: 0.25–0.5 mg/kg divided twice daily Max 12 mg/24 h 2–6 years of age Max 16 mg/24 h 7–14 years of age	Placebo ($n=14$)	**Self-reported change of frequency and duration of abdominal pain** **Parents' impression or assessment of the children's improvement**	**Self-reported change of frequency and duration of abdominal pain, n (%)**	
				Frequency *Cyproheptadine* Completely resolved: 3 (20 %) Very much improved: 6 (40 %) Improved: 4 (26.7 %) *Placebo* Completely resolved: 0 (0 %) Very much improved: 2 (14.2 %) Improved: 3 (21.4 %)	$p=0.002$
				Intensity *Cyproheptadine* Completely resolved: 3 (20 %) Very much improved: 6 (40 %) Improved: 4 (26.7 %) *Placebo* Completely resolved: 0 (0 %) Very much improved: 2 (14.2 %) Improved: 2 (14.2 %)	$p=0.001$
				Parents' impression or assessment of the children's improvement, *n* (%) *Cyproheptadine* Satisfactory: 13 (86.7 %) Unsatisfactory: 2 (13.3 %) *Placebo* Satisfactory: 4 (28.6 %) Unsatisfactory: (71.4 %)	$p=0.003$

Studies Evaluating Medications in Recurrent Abdominal Migraine (AM)

Medication	Intervention	Control intervention	Outcome measured	Results	Efficacy
Propranolol, cyproheptadine [40]	Retrospective review of patients with abdominal migraine that underwent treatment ($n=53$) Propranolol: dose not discussed Cyproheptadine: dose not discussed	None	**Response to treatment** Excellent: cessation of abdominal pain Fair: persistence of symptoms but milder and less frequent Poor: no response	**Response to treatment** *Propranolol* ($n=24$) Excellent: 18/24 (75 %) Fair: 2/24 (8 %) Poor: 4/24 (17 %)	*p*: not significant (trend toward favoring propranolol)
				Cyproheptadine ($n=12$) Excellent: 4/12 (33 %) Fair: 6/12 (50 %) Poor: 2/12 (17 %) (Note: Four patients did not receive treatment)	Patients with abdominal migraine may benefit from prophylactic treatment with propranolol or cyproheptadine
				Average number abdominal pain attacks (mean ± standard error) *Propranolol* 6 months before treatment: 5.4 ± 0.6 6 months after treatment: 1.0 ± 0.3 *Cyproheptadine* 6 months before treatment: 5.8 ± 2.1 6 months after treatment: 1.3 ± 0.5	
Valproic acid [41]	Case reports of patients with abdominal migraine refractory to other therapy were given valproic acid ($n=2$) Case 1: valproic acid 500 mg intravenously three times daily at each admission titrated to serum levels 100–120 mcg/dL and converted to oral therapy to continue out patient Previous medications *No response* metoclopramide, ondansetron, promethazine *Mild response* Sumatriptan Dihydroergotamine Risperidone Haloperidol Chlorpromazine Lorazepam Diphenhydramine Case 2: valproic acid intravenously (unknown dose) Previous medications Lorazepam Haloperidol Olanzapine	None	**Symptom resolution**	**Symptom resolution** Case 1: rapid resolution of symptoms with valproate administration (Medication discontinued as outpatient due to weight gain, continued to use with success for acute episodes) Case 2: pain symptoms began to improve as valproic acid serum levels became therapeutic	Intravenous valproic acid has potential to resolve pain and mental status changes in patients with abdominal migraine

Studies Evaluating Medications in Aerophagia

Medication	Intervention	Control intervention	Outcome measured	Results	Efficacy
Omeprazole ($n=1$), Aluminum hydroxide ($n=2$), Simethicone ($n=4$), Chlordiazepoxide ($n=2$), Metoclopramide ($n=1$), Dicyclomine ($n=1$) [43[a]]	Retrospective chart review of various treatments: education ($n=32$), behavioral therapy ($n=4$), medications ($n=9$)	None, retrospective review	**Symptom improvement** Documented follow-up visit ($n=12$) with improvement in symptoms, not worsened, no recurrent visit for aerophagia	**Symptom improvement:** Improvement in symptoms, not worsened, no recurrent visit for aerophagia: $n=4$ Of patients that reported improvement: $n=1$ treated with medications (chlordiazepoxide, aluminum hydroxide, simethicone) $n=3$ education alone	N/A
Clonazepam [44]	Retrospective review with 1-year follow-up Clonazepam ($n=15$): <30 kg: initial 0.025 mg/kg/day in 2 divided doses, increased by 0.025 mg/kg every 3–5 days as needed, maintenance usually 0.1 mg/kg >30 kg: initial and maintenance 1.5 mg daily in two divided doses	Reassurance ($n=7$), parents chose to be in treatment or control group	**Positive response**: symptom-free for 1 consecutive week within 1 month of treatment **Remission**: symptom-free for 1 consecutive month within 6 months treatment	**Positive response:** Clonazepam: 40 % Reassurance: 0 %	$p=0.067$
				Remission: Clonazepam: 66.7 % Reassurance: 14.3 %	$p=0.032$
Osmotic laxatives (milk of magnesia ($n=2$), polyethylene glycol ($n=5$)) [45]	Prospective, observational study Increase laxative dose Discontinued osmotic laxative and switched to stimulant ($n=5$) (senna or bisacodyl)	None, observational study Laxatives given until diagnosis of aerophagia then reassurance, education, behavioral modifications used as treatment	**Resolution of aerophagia**	**Resolution of aerophagia:** All patients at 2–20 months (mean = 7.9 months)	N/A

[a]*Note*: Patients may have been started on multiple medications, and total *n* started on medications and *n* that received medication intervention are not the same

Studies Evaluating Medications in Cyclic Vomiting Syndrome (CVS)

Medication	Intervention	Control intervention	Outcome measured	Results	Efficacy
Treatment					
Erythromycin [19]	Prospective, case series of patients with cyclic vomiting ($n=21$) refractory to metoclopramide ($n=13$) were given erythromycin for acute management after diagnostic evaluation Erythromycin ethylsuccinate: 20 mg/kg/day in 2–4 divided doses for 7 days	None	**Emesis at follow-up** (initial, 2 and 6 months post-intervention): Responders Non-responders 50 % improvement	**Initial response** Responders, complete: 11/21 Non-responders: 7/21 50 % improvement: 2/21 **2 and 6 month response** Responders: 13/21 (no further emesis) Note: Two patients discontinued therapy due to reported side effects (vomiting or diarrhea)	N/A
Erythromycin [20]	Prospective, case series of patients with cyclic vomiting ($n=24$) refractory to metoclopramide ($n=14$) were given erythromycin for acute management after diagnostic evaluation Erythromycin ethylsuccinate: 20 mg/kg/day in 2–4 divided doses for 7 days	None	**Emesis at follow-up** (Initial, 2,6,12,24 months post-intervention): Responders Non-responders 50 % improvement	**Initial response** Responders, complete: 14/24 Non-responders: 6/24 50 % improvement: 2/24 **2, 6, 12, 24 month response** Responders: 18/24 (no further emesis) Note: Four patients discontinued therapy due to reported side effects (vomiting or diarrhea)	N/A
Promethazine, ondansetron, oxazepam (other benzodiazepines) [21]	Case report of patient (12-year-old) with cyclic vomiting syndrome with a 7-year history of episodes treated as described for acute episodes ($n=1$) (only medications mentioned, treatment included multimodal approach) First episode: oxazepam 10 mg, promethazine 50 mg First recurrence: Ondansetron 8 mg intravenously Second recurrence: ondansetron 4 mg	None	**Vomiting episodes**	**Vomiting episodes** Lessened in frequency and intensity 18 months of follow-up Patient began attending school, gained weight, and progressed emotionally and physically	Improvement with multimodal approach

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Medication	Intervention	Control intervention	Outcome measured	Results	Efficacy
Barbiturates (phenobarbital) [22]	Case series of patients with cyclic vomiting syndrome given phenobarbital for acute management ($n=14$) Phenobarbital: 30–120 mg/h (mean 2 mg/kg/day, median dose 60 mg/h) Previous therapy ineffective: Propranolol ($n=3$) Butalbital (n-2)	None	**Symptom resolution**	**Symptom resolution** 11/14 complete resolution 3/14 marked improvement	Improvement in all patients noted
Dexmedetomidine [23]	Case reports of patients with cyclic vomiting syndrome given dexmedetomidine for acute management ($n=3$) unresponsive to home regimens Dexmedetomidine: Bolus 0.25–0.5 mcg/kg Continuous infusion 0.25 mcg/kg/h × 12–18 h	None	**Symptom resolution**	**Symptom resolution** Effective 4/5 episodes Case 1: immediate improvement with 2 courses during acute episode, no response one episode Case 2: immediate improvement with 1 course during acute episode Case 3: Improvement with 1 course during acute episode	Improvements noted in all patients
Sumatriptan [24]	Case report of a patient with cyclic vomiting syndrome after many hospital admissions refractory to many therapies was given sumatriptan for acute management Sumatriptan: 20 mg intranasal	None	**Symptom resolution**	**Symptom resolution** Complete improvement "soon" after administration (typical episodes last ≥24 h)	Improvement noted for acute treatment
Sumatriptan [25]	Prospective, non-blinded, non-placebo-controlled study of patients with cyclic vomiting were given sumatriptan for acute management ($n=12$) Sumatriptan: dose [(age x4 + 20)/100 × 3 mg] subcutaneously or 20 mg nasally	None	**Response** Complete: no vomiting after treatment Effective: frequency of vomiting reduced by at least half compared to last attack Non-effective: not effective in preventing vomiting	**Response (n, patients)** Complete: 4/12 Effective: 5/12 Non-effective: 3/12 **Response (n, attack)** 19/35 (54 %)	Improvement noted in 75 % patients for acute treatment

Clonidine [26]	Case report of a patient with cyclic vomiting syndrome refractory to various medications was given clonidine for acute management on various admissions Clonidine: 25 mcg (1 mcg/kg) intravenously then 50 mcg (2 mcg/kg) intravenously (various doses given different admissions) repeated every 6–8 h according to symptoms Other therapies: ondansetron, cyproheptadine, pizotifen, sumatriptan, chlorpromazine, benztropine, omeprazole, dexamethasone, amitriptyline	None	**Symptom resolution**	**Symptom resolution** 6/6 episodes (continue to use as routine management for patient during acute episodes)	Improvement noted for acute treatment
Prophylaxis					
Topiramate [35]	Case report of a patient with cyclic vomiting syndrome and generalized epileptiform discharges refractory to multiple therapies was given topiramate (for epilepsy) Topiramate: 3 mg/kg/day orally and titrated to 5 mg/kg/day Other therapies: "anti-ulcer," "anti-reflux," cyproheptadine, flunarizine	None	**Symptom resolution**	**Symptom resolution** Symptoms resolved after initiation with no further attacks × 1 year, returned once medication withdrawn and resolved with medication reinitiation	Improvement noted for prophylactic treatment
Propranolol, pizotifen, ergot, cyproheptadine, "antiepileptics," "antidepressant," multivitamin, "homeopathic" [16]	Retrospective review conducted through means of a questionnaire sent to families of patients with cyclic vomiting syndrome ($n = 31$) that had taken medications for prophylaxis Patients reporting use of prophylactic medication ($n = 11$)	None	**Reported benefit** Family perception of success to therapy, clinical and psychological features, precipitants, interventions utilized	**Reported benefit** Propranolol: 4/6 Antimigraine (pizotifen, ergot, cyproheptadine): 2/7 Antiepileptics: 0/3 Antidepressant: 0/1 Homeopathic and multivitamins: 3/6 **Benefit from prophylactic therapy more likely to benefit those with:** Less episodes of vomiting per year Less intense episodes Episode precipitated by infection Those with concomitant migraine	Certain patients may benefit from prophylactic therapy

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Medication	Intervention	Control intervention	Outcome measured	Results	Efficacy
Cyproheptadine, amtriptyline [28]	Retrospective chart review of patients with cyclic vomiting syndrome treated with prophylactic amitriptyline or cyproheptadine ($n=27$) with: Amitriptyline: initially 10 mg orally at bedtime, titrated to effect or adverse effects (range 10–200 mg/day, 0.2–3.4 mg/kg/day) Cyproheptadine: initially 2 mg orally twice daily, titrated to effect or adverse effects (range 4–12 mg/day, 0.1–0.3 mg/kg/day) Previous therapy ineffective: Antiemetics, prokinetic agents, antibiotics, H2-blockers, antisecretory agents, anticholinergics, antidepressants, anticonvulsants, and antacids	None	**Response** Complete: no attacks Partial response: ≥50 % reduction in frequency of attacks No response: <50 % decrease in frequency of attacks	**Response** *Amitriptyline* Complete: 16/22 (73 %) Partial: 4/22 (18 %) No response: 2/22 (9 %) *Cyproheptadine* Complete: 4/6 (66 %) Partial: 1/6 (17 %) No response: 1/6 (17 %) **Follow-up** *Amitriptyline* Mean: 38.1 months *Cyproheptadine* Mean: 11.7 months	Both agents were effective for prophylaxis
Amitriptyline [29]	Case report of a patient with cyclic vomiting syndrome that was given amitriptyline ($n=1$) Other therapy: ondansetron	None	**Response** Acute episodes	**Response** 2 acute episodes in 18 months (reduced from 7 to 10 episodes per year prior to therapy initiation)	Benefit noted for this patient
Nebivolol, amitriptyline [30]	Case report of a patient with cyclic vomiting syndrome that was given nebivolol and amitriptyline ($n=1$) Amitriptyline: 25 mg orally daily Nebivolol: unknown dose Other therapy: ondansetron Previous therapy ineffective: antisecretory agent, topiramate, valproate	None	**Response** Acute episodes	**Response** 1 acute episode in 6 months (reduced from a peak of 1 episode per month prior to therapy initiation)	Benefit noted for this patient
Amitriptyline, coenzyme q-10 [31]	Retrospective review conducted through means of a questionnaire sent to families of patients with cyclic vomiting syndrome ($n=130$) that had taken amitriptyline ($n=113$) or coenzyme q-10 ($n=17$) Amitriptyline Coenzyme q-10 Note: Adults were also included in this study	None	**Efficacy** (compound definition based on vomiting frequency, episode duration, number of emeses, nausea severity) Positive: ≥50 % reduction in each parameter Negative: <50 % reduction in each parameter	**Efficacy** Amitriptyline: 72 % Coenzyme q-10: 69 %	Odds ratio 1.2 (95 % CI 0.5–3.0)
				Adverse effects Amitriptyline: 50 % Coenzyme q-10	$p=5\times10^{-7}$
				Amitriptyline: 21 % discontinuation Coenzyme q-10: 0 % discontinuation	$p=0.007$
				Patient satisfaction Amitriptyline: 47 % Coenzyme q-10: 77 %	$p=0.008$ Odds ration 3.6 (95 % CI 1.2–10)

Amitriptyline, coenzyme q-10, L-carnitine, topiramate [32]	Retrospective, case series, chart review of patients with cyclic vomiting syndrome that were given a combination of dietary recommendations, coenzyme q-10, L-carnitine, amitriptyline, or cyproheptadine, topiramate ($n=30$) Coenzyme q-10: initial 10 mg/kg/day orally or 100 mg orally twice daily (whichever was smaller) L-carnitine: initial 100 mg/kg/day orally divided twice daily or 2 g orally twice daily (whichever was smaller) Amitriptyline: ≥ 5 years initial 0.5 mg/kg/day orally at night Cyproheptadine: <5 years initial 0.25 mg/kg/day orally divided twice a day Topiramate: 25 mg orally twice daily ($n=2$) Doses were adjusted based on serum levels and study specified maximum doses	None	**Vomiting episodes** **Response** **Adverse effects**	**Vomiting episodes** Resolved 23/30 Improved by >75 % 3/30 Improved by >50 % 1/30 Failure 3/30 **Response** 26/29 (>75 %) **Adverse effects** Amitriptyline $n=9/19$ Coenzyme q-10 $n=1$ Cyproheptadine $n=1$ Multiple medications $n=2$	Substantial efficacy noted with protocol
Valproate, phenobarbital [37]	Case reports of patients with cyclic vomiting syndrome refractory to multiple therapies were given valproate and phenobarbital ($n=2$) Valproate sodium: 20–26 mg/kg/day Phenobarbital: 4–5 mg/kg/day Case 1: refractory to amitriptyline, carbamazepine, phenytoin, cyproheptadine, valproate, and phenobarbital monotherapy. Given phenobarbital and valproate in combination Case 2: refractory to diazepam, phenytoin, valproate, and phenobarbital monotherapy. Given phenobarbital and valproate in combination	None	**Response to combination therapy**	**Response to combination therapy** Both had cessation of vomiting episodes	Effective in these two patients

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Medication	Intervention	Control intervention	Outcome measured	Results	Efficacy
Cyproheptadine, amitriptyline, propranolol [33]	Retrospective chart review of patients admitted to the hospital and diagnosed with cyclic vomiting syndrome with follow-up in an outpatient clinic ($n=24$) Amitriptyline: unknown doses Cyproheptadine: unknown doses Propranolol: unknown doses	None	**Symptom improvement** Frequency of acute episodes Duration of acute episodes	**Severe episodes (n)** Prior to therapy: average 2.3 ± 2.0 per year After therapy initiation: Average 0.6 ± 1.1 per year **Duration of vomiting episode (days)** Average 4.1 ± 2.2 (range: 1.5–10) Average 0.2 ± 1.4 18/24 patients had improvement in duration and frequency of episodes after starting therapy (4 lost to follow-up and 2 sought alternative therapy)	Improvement of duration and frequency of episodes with cyproheptadine and/or amitriptyline as prophylaxis
Amitriptyline, pizotifen, propranolol [17]	Retrospective chart review of patients with cyclic vomiting syndrome evaluated for data including treatment and outcome ($n=25$) All patients received prophylactic regimens for at least 3 months	None	**Response** Good: absence or few episodes of vomiting Fair: persistence of vomiting but improvement with less frequency and less intense episodes Poor: no response	**Response** *Amitriptyline* Good: 11/18 Fair: 4/18 Poor: 3/18 *Pizotifen* Good: 3/8 Fair: 1/8 Poor: 4/8 *Propranolol* Good: 1/2 Fair: 0/2 Poor: 1/2	Amitriptyline and pizotifen are effective in prophylactic therapy
				Comparison of Response Amitriptyline (83.3 %) vs. pizotifen (50 %)	$p=0.14$
Amitriptyline, propranolol [34]	Prospective, randomized study of patients with cyclic vomiting syndrome evaluated, treated, and followed ($n=181$) Amitriptyline: initial 1 mg/kg/day orally Propranolol: initial 1 mg/kg/day orally	None	**Efficacy** Not defined	**Efficacy**	
				Amitriptyline: 46/81 (56 %)	$p<0.001$
				Propranolol: 74/83 (92 %)	$p<0.0001$
				Adverse effects Amitriptyline: irritability, agitation, insomnia, or lethargy Propranolol: no significant adverse effects	Propranolol appears more effective than amitriptyline for prophylactic use

Valproate, phenobarbital [38]	Prospective, nonrandomized study of patients with cyclic vomiting syndrome given prophylactic therapy with valproate ($n=13$) ± phenobarbital ($n=3$) Valproate: initial 10 mg/kg/day orally in 2 divided doses titrated to response (10–40 mg/kg/day) Phenobarbital: added if unresponsive to monotherapy 4–5 mg/kg/day Previous therapy ineffective: amitriptyline including irritability, agitation, insomnia, or lethargy	None	**Response** Complete: no vomiting Marked: <2 attacks per year None: no change in frequency of attacks	**Response** Complete: 2/13 Marked: 9/13 None: 2/13 85 % response rate (complete and marked total, 3 patients on concomitant therapy included in this value)	Valproate appears to be effective for the prophylactic management of severe CVS
L-carnitine [36]	Case reports of patients with cyclic vomiting syndrome that were given L-carnitine ($n=6$) L-carnitine: 30–150 mg/kg/day orally	None	**Time between episodes**	**Time between episodes** Prior to therapy: average 1.7 months After therapy initiation: Average 1.1 years	Oral L-carnitine may be useful to control the symptoms in patients with CVS
Treatment and prophylaxis					
Ondansetron, erythromycin, propranolol, cisapride, pizotifen [18]	Retrospective review and interview of patients with cyclic vomiting syndrome followed by clinic ($n=6$) All: doses not discussed	None	**Response** **Duration of attack and amount of emesis (acute)**	**Response** **Duration of attack and amount of emesis (acute)** Ondansetron: 1/3 noted shorter duration and fewer emeses (not responsive on subsequent episodes) Erythromycin: 0/1 no response	Response to both acute and prophylactic treatment was disappointing
			Reduction of episodes (prophylaxis)	**Reduction of episodes (prophylaxis)** Propranolol: 0/4 no reduction in episodes Cisapride: 0/2 no reduction in episodes Pizotifen: 0/2 no reduction in episodes	

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Medication	Intervention	Control intervention	Outcome measured	Results	Efficacy
Amitriptyline, cyproheptadine, propranolol, ondansetron, lorazepam, promethazine, sumatriptan [118]	Retrospective review with distribution of questionnaire surveying patients with cyclic vomiting syndrome assessing if the presence of neuromuscular manifestations differentiates this as a subgroup of CVS	None	**Patient reported benefit**	**61 different medications used and 54/61 considered efficacious** **Patient reported significant benefit, *n/N*, (%)** Amitriptyline 16/31 (52 %) Cyproheptadine 11/18 (61 %) Propranolol 5/15 (33 %) Ondansetron 42/52 (81 %) Lorazepam 23/30 (77 %) Promethazine 13/23 (57 %) Sumatriptan 8/8 (100 %)	Our methods were anecdotal and the small numbers reported on each agent do not allow for a careful evaluation of the efficacy of various treatment modalities, especially any comparison between our subject groups or between therapeutic agent
Amitriptyline, cyproheptadine, propranolol, ondansetron, lorazepam (sedatives), promethazine, sumatriptan, anticonvulsants, caffeine, dexamethsone [119]	Retrospective review with clinical interview through questionnaire of patients with CVS + (cyclic vomiting syndrome with neuromuscular disease manifestations) that were given multiple treatments as part of their course ($n = 62$)	None	**Positive response** (as reported by patients or caregivers, positive response not defined)	**Positive response (*n/N*, %)** *Prophylactic* Amitriptyline: 17/22 (77 %) Cyproheptadine: 8/14 (57 %) Propranolol: 6/13 (46 %) (Note: Three patients discontinued amitriptyline due to adverse effects) *Abortive/supportive* Ondansetron: 24/52 (46 %) ≥2/8 lorazepam/sedation ≥2/7 promethazine *n* unknown (≥2): sumatriptan, anticonvulsants, caffeine, dexamethasone	In most cases antimigraine (i.e., amitriptyline) in combination with anti-mitochondrial/ metabolic therapy was highly effective

Promethazine, cisapride, ondansetron, isometheptene, propranolol, cyproheptadine, amitriptyline, sumatriptan [120]	Retrospective chart review and structured interviews of patients with cyclic vomiting syndrome ($n=214$) ± migraine-related symptoms, acute and chronic management was evaluated ($n=176$)	Non-migraine-associated cyclic vomiting syndrome ($n=38$)	**Response to medications** (Percent having a >50 % reduction in vomiting for abortive therapy or in number of episodes for prophylactic therapy)	**Response to medications**	
				Promethazine Migraine-associated CVS 23 %, $n=48$ Non-migraine-associated CVS 13 %, $n=15$	$p=0.423$
				Cisapride Migraine-associated CVS 19 %, $n=32$ Non-migraine-associated CVS 25 %, $n=8$	$p=0.693$
				Ondansetron Migraine-associated CVS 77 %, $n=35$ Non-migraine-associated CVS 71 %, $n=7$	$p=0.746$
				Isometheptene Migraine-associated CVS 31 %, $n=13$ Non-migraine-associated CVS 0 %, $n=0$	N/A
				Propranolol Migraine-associated CVS 71 %, $n=52$ Non-migraine-associated CVS 38 % $n=8$	$p=0.060$
				Cyproheptadine Migraine-associated CVS 47 %, $n=32$ Non-migraine-associated CVS 0 %, $n=5$	$p=0.047$
				Amitriptyline Migraine-associated CVS 75 %, $n=12$ Non-migraine-associated CVS 100 %, $n=1$	$p=0.569$
				Sumatriptan Migraine-associated CVS 69 %, $n=35$ Non-migraine-associated CVS 33 %, $n=3$	$p=0.217$
				Any migraine therapy Migraine-associated CVS 79 %, $n=86$ Non-migraine-associated CVS 36 %, $n=11$	$p=0.002$

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Medication	Intervention	Control intervention	Outcome measured	Results	Efficacy
Midazolam, clonidine [27]	Case report of a patient with cyclic vomiting syndrome was given midazolam and clonidine for acute and chronic management ($n=1$, 54 kg) *First episode* Midazolam: 1 mg/h intravenously weaned over 36 h with initiation of oral clonidine Clonidine: 2 mcg/kg (100 mcg) intravenously then 100 mcg twice daily orally ×6 days *Second episode* Midazolam: 1 mg/h intravenously increased to 2 mg/h then weaned after initiation of oral clonidine Clonidine: 2 mcg/kg (100 mcg) intravenously then discharged on 100 mcg twice daily orally Home therapy prior to admission: prophylactic pizotifen 1.5 mg nightly and acute ondansetron (4 mg intravenously day 1 and 8 mg three times daily on day 2) with persistent symptoms	None	**Symptom resolution**	**Symptom resolution** Combination of midazolam and clonidine showed dramatic improvement over 12–36 h in 2 episodes	Improvement noted for acute and prophylactic management
			Number of acute episodes	**Number of acute episodes** 2 acute episodes in 2 years	

Studies Evaluating Medications in Encopresis

Medication	Intervention	Control intervention	Outcome measured	Results	Efficacy
Lactulose [53]	Prospective, randomized, non-blinded study × 7 weeks of treatment (12 month follow-up)	Biofeedback ($n=25$)	**Reduction of encopresis episodes/week**	**Reduction of encopresis episodes/week** BF + LAX = 2 BF = 2	$p<0.05$ both groups
	Biofeedback + lactulose ($n=23$) 5g/10 kg/day divided into 2 doses stopped when: 1. <2 encopresis episodes/month or 2. Increase in encopresis episodes		**Colonic transit time**	**Colonic transit time** BF + LAX = 34 h, median BF = 34 h, median	No difference
			Anorectal manometry, normal defecation dynamics	**Anorectal manometry, normal defecation dynamics** BF + LAX = change 15 % BF = change 13 %	No difference
Milk of magnesia Senna "enema/ suppository" [54]	Prospective, randomized study with 12-month follow-up/ treatment Intense medical therapy ($n=29$) Intense medical therapy + behavior management with enhance toilet training ($n=27$) Intense medical therapy + behavior management with enhance toilet training + biofeedback therapy ($n=31$) IMT: 1. Disimpaction with enemas 2. Laxatives titrated to 1–3 soft stools per day 3. Enema or suppository if no bowel movement in 48 h	None	**Daily frequency of fecal soiling at 3, 6, and 12 months** **Number of bowel movements in toilet per day** **Frequency of self-initiated toileting per day** **Average amount of laxative used per day**	**Decrease in average daily frequency soiling**: IMT = 0.68 at 3 months IMT = 0.52 at 6 months IMT = 0.48 at 12 months ETT = 0.21 at 3 months ETT = 0.45 at 6 months ETT = 0.53 at 12 months BF = 0.51 at 3 months BF = 0.26 at 6 months BF = 0.37 at 12 months	Significant improvement in all groups, no statistically significant difference among groups ($p>0.2$): All regimens were associated with significant and sustained decrease in average daily frequency of soiling

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Medication	Intervention	Control intervention	Outcome measured	Results	Efficacy
				Number of bowel movements in toilet per day: IMT baseline = 0.97 IMT = 1.44 at 3 months IMT = 1.36 at 6 months IMT = 1.30 at 12 months ETT baseline = 0.84 ETT = 1.21 at 3 months ETT = 1.31 at 6 months ETT = 1.01 at 12 months BF baseline = 0.96 BF = 1.25 at 3 months BF = 1.12 at 6 months BF = 1.16 at 12 months	Significant improvement in all groups, no statistically significant difference among groups ($p > 0.2$): All regimens associated with increased frequency of bowel movements passed in toilet
				Frequency of self-initiated toileting per day: IMT baseline = 1.25 IMT = 1.53 at 3 months IMT = 1.49 at 6 months IMT = 1.40 at 12 months ETT baseline = 1.41 ETT = 1.62 at 3 months ETT = 1.67 at 6 months ETT = 1.31 at 12 months BF baseline = 1.32 BF = 1.40 at 3 months BF = 1.34 at 6 months BF = 1.31 at 12 months	No significant increases in any group, no statistically significant difference among groups ($p = 0.7497$)

				Average amount of laxative used per day at 12 months: IMT = 11.7 mL/day ETT = 3.7 mL/day BF = 9.25 mL/day	Statistically significant difference among groups ($p = 0.04$)
				Total cure (no fecal soiling during 2-week assessment period) at 12 months: IMT 34.5 % ETT 44.4 % VF 35.5 %	No statistically significant difference among groups ($p = 0.7005$)
Erythromycin [121]	Prospective study of patients with colonic motility disorders ($n = 20$, $n = 12$ constipation/encopresis) Erythromycin: 3 mg/kg intravenously by slow push	None	**Manometry**: measured high-amplitude propagating contractions (HAPC) (≥60mmHg in amplitude, 10 s in duration, propagating for at least 30cm of the colon)	3/14 healthy patients HAPC's post-erythromycin Motility index before and after erythromycin infusion	No changes in colonic motility index
				Manometry 60 min before: 254 ± 74 mmHg/h 60 min after: 253 ± 94 mmHg/h	$p = 0.55$
				15 min before: 64 ± 23 mmHg/15 min 15 min after: 69 ± 32 mmHg/15 min	$p = 0.45$

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Medication	Intervention	Control intervention	Outcome measured	Results	Efficacy
Milk of magnesia, lactulose, sorbitol, mineral oil, senna [122]	Prospective study of patients with functional encopresis (symptoms of functional fecal retention and those matching diagnosis according to Rome II criteria) identified at clinic visit ($n=213$) Initial: milk of magnesia 1mL/kg/day in patients with rectal fecal mass and 2.5mL/kg/day in patients with abdominal fecal mass Adjustment: 1. Milk of magnesium titrated to 1–2 soft bowel movements per day without fecal soiling or abdominal pain 2. Lactulose, sorbitol, mineral oil used if milk of magnesia refused 3. Senna given if no response to laxatives	None	1-year assessment per patient interview/ questionnaire assessing **successful treatment and recovery:** Successful treatment: ≥3 bowel movements/ week, no soiling, no abdominal pain, independent of laxative use Recovered: ≥3 bowel movements/week and ≤2 smears/month, while off laxatives for 1 month	**Functional fecal retention by:** Symptoms and physical exam ($n=181$) Successful treatment: 50 % Recovered: 39 % **Rome II criteria ($n=88$):** Successful treatment: 51 % Recovered: 39 %	N/A: focus was to identify issues with diagnosing patients with functional fecal retention according to Rome II criteria
Laxatives [123]	Retrospective chart review identifying children with fecal incontinence and no underlying cause that underwent anorectal manometry ($n=13$) Treatment: combination of education, dietary modification, stimulatory laxatives ($n=2$)	Controls with constipation ($n=11$)	**Improved soiling** 6-month treatment period with report Anorectal manometry characteristics (not reported here)	**Improved soiling** Decreased fecal soiling (6/8) 2/2 treated with laxatives 2/4 treated with dietary modification 2/2 treated with education	Improved in both patients treated with laxatives
Laxatives [124]	Randomized, parallel prospective study evaluating treatment of patients with fecal incontinence given medical management (MM) with behavior modification ($n=83$) Initial: disimpaction with up to 4 cycles: 1. Enema 5mL rectally (90 mg sodium abate, 9 mg sodium lauryl sulfoacetate, 5 mg sorbic acid, glycerol, sorbitol, distilled water) on day 1 2. Bisacodyl 5 mg rectally ×2 on day 2 3. Bisacodyl 5 mg orally ×2 on day 3 Maintenance: liquid paraffin 5–30mL once to twice daily, senna granules and/or bisacodyl tablets. Medications were adjusted to maintain daily defecation	Behavior modification (BM) only ($n=86$)	**Remission at 3, 6 and 12 months** Full Full and partial remission	**Remission** 3 months MM: 39 % full BM: 12 % full MM: 8 % partial BM: 12 % partial 6 months MM: 49 % full BM: 30 % full MM: 11 % partial BM: 6 % partial 12 months MM: 51 % full BM: 36 % full MM: 12 % partial BM: 7 % partial	All p values significant except* difference between MM and BM group full response at 12 months Note: p values for partial remission reported as combination of full and partial (listed here as partial only) Laxative treatment combined with behavior modification for encopresis is superior for remission/ improvement compared to behavior modification alone

Medication	Intervention	Control intervention	Outcome measured	Results	Efficacy
Multimodal [125]	Retrospective analysis of patients with encopresis at presentation to clinic for treatment and analysis at 1 year Initial: "Cleanout" with four cycles 1. "Enema" rectally on day 1 2. "Suppository" rectally on day 2 3. "Laxative tablet" orally on day 3 Maintenance: Multimodal approach individualized for each patient to maintain regular stools and no soiling (details not specified) 1. Mineral oil and/or other laxatives and stool softeners 2. Toilet retraining 3. Dietary recommendations	None	**Short-term improvement** (2-week follow-up)	**Short-term improvement** 30/45 significantly improved (no soiling outside of intervention induced soiling)	Unable to attribute improvement to medications due to study design and manner in which outcomes were reported
			Compliance	**Compliance** Good: 29/45 Fair: 7/45 Poor: 3/45	
			Long-term follow-up (mean duration 53 months, range 15–99 months)	**Long-term follow-up** 26/45 (58 %) no soiling or <1/month 13/45 (29 %) soiling ≥1/month but <initial presentation 6/45 (13 %) soiling ≥1/month with frequencies ≥initial presentation	
Laxatives, senna [126]	Retrospective case review and analysis of patients with fecal incontinence ($n=78$) followed by treatment of patients with fecal incontinence ($n=59$) according to author's defined group classification Group 1: *training problems without severe constipation* 2/10 given laxatives for *mild* constipation Group 2: *pot refusal retention syndrome* 19/19 given oral laxatives soften stool and increase urge to defecate • Senna at various doses (cleanout followed by maintenance) Group 3: *severe constipation with overflow* 19/19 given oral laxatives to soften stool • Senna at various doses (cleanout followed by maintenance) Group 4: *uncomplicated functional fecal incontinence* 0/11 given laxatives	None	**Remission at 6 months**	**Remission** Group 1: 8/10 (unknown if included patients given laxatives) Group 2: 17/19 Group 3: 16/19 Group 4: 1/11 Severely constipated soilers: 33/38 (87 %)	Use of laxatives with behavioral management supported in patients with severe constipation

BF biofeedback, *LAX* laxative, *IMT* intense medical therapy, *ETT* enhanced toilet training, *mL* milliliters

Studies Evaluating Medications in Functional Dyspepsia

Medication	Intervention	Control intervention	Outcome measured	Results	Efficacy
Famotidine [55]	Prospective, blinded, crossover study ×3 weeks treatment and 12-month follow-up 0.5 mg/kg/dose twice daily (max 40 mg/day) for 3 weeks with crossover when symptoms recurred or at the end of treatment period ($n=25$)	Placebo	**Quantitative overall score** Abdominal pain score (APS) Pain frequency score Pain severity score Peptic index score	**Quantitative overall score** Change in APS: Famotidine = 3.37 ± 3.53 Placebo = 1.66 ± 2.7	Not significant ($p=0.16$)
			Global assessment Better Not better Worse	**Global assessment** 68 % improved with famotidine vs. 12 % with placebo	Statistically significant
Amitriptyline [56]	Prospective, double-blind, randomized, placebo-controlled parallel-group study ×4 weeks treatment following 1 week observational period Amitriptyline ($n=43$) 10 mg/day <35 kg, 20 mg/d > 35 kg ×4 weeks in patients with irritable bowel syndrome, functional abdominal pain or **functional dyspepsia**	Placebo ($n=40$)	**Overall response to treatment** (self-assessment of pain relief and sense of improvement) **Effect on psychosocial traits** **Effect on daily functioning**	**Overall response to treatment** Self-reported feeling better: Amitriptyline: 63 % Placebo: 57.5 %	Not significant ($p=0.63$)

Tricyclic antidepressant (TCA) [11]	Retrospective review of patients prescribed amitriptyline or imipramine ($n=146$) for irritable bowel syndrome, **dyspepsia**, functional disorder of the intestine	N/A	**Overall response to treatment**	**Overall response to treatment** Responded: 78.6 % for average of 10.73 months Amitriptyline Responded well: $n=40$ (FD $n=4$) Did not respond: $n=8$ (FD $n=2$) Stopped due to adverse effects: $n=3$ (FD $n=0$) Imipramine Responded well: $n=41$ (FD $n=8$) Did not respond: $n=5$ (FD $n=2$) Stopped d/t adverse effects: $n=2$ (FD $n=0$)	TCAs effective in treating function gastrointestinal disorders in pediatric patients
Lactobacillus rhamnosus GG [57]	Meta-analysis evaluating utility of *lactobacillus GG* in functional disorders (one study included functional dyspepsia, $n=20$) 3×10^9 CFU twice daily for 4 weeks	Placebo	**Treatment success** No pain at the end of the intervention	**Treatment success** Relative risk reduction for abdominal pain for functional dyspepsia in one trial = 0.83	No difference
				Frequency of pain	No difference
				Pain intensity	No difference
Rifaximin [10]	Prospective, double-blind, placebo-controlled study of patients with **functional dyspepsia** ($n=11$), IBS ($n=41$), functional abdominal pain ($n=23$), or abdominal migraine ($n=0$) that were given rifaximin ($n=49$) Rifaximin: 550 mg orally three times daily × 10 days	Placebo ($n=26$)	**Symptom improvement (overall and individual symptoms)** **Hydrogen levels** (not reported here) **Methane levels** (not reported here)	**Symptom improvement** Specifics not mentioned in the study	All values not significant between groups

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Medication	Intervention	Control intervention	Outcome measured	Results	Efficacy
Montelukast [58]	Double-blind, placebo-controlled, randomized, crossover study of patients with functional dyspepsia and duodenal eosinophilia ($n=40$) Montelukast: 10 mg orally daily × 14 days (note: all patients also took ranitidine, except two patients)	Placebo	**Symptomatic response** Patient diaries Five point global pain relief evaluation: • Grade 1: worse • Grade 2: no change • Grade 3: moderate improvement (partial response, improved pain) • Grade 4: good (nearly complete relief, pain does not interfere with daily activities) • Grade 5: excellent (complete relief of pain) **Biochemical response** Eosinophil count **Data not reported here** Complete blood count Serum IgE Others Montelukast serum levels	**Symptomatic response** *Relief in pain grades 3–5 ("responders") in first 14 days* Montelukast: 62.1 % Placebo: 32.4 % *Relief in pain grades 3–5 at crossover* Montelukast to placebo: 45 % (38 % had increase in pain) Placebo to montelukast: 20 % (62 % had increase in pain)	$p<0.02$
				Eosinophil count Patients with peak eosinophil counts >20/high power field ($n=31$) "responder" rate to montelukast 68 % vs. 32 % placebo	$p=0.01$ Appears to be a role for montelukast in pediatric patients between 7 and 17 years of age with dyspepsia and duodenal eosinophilia defined as peak eosinophil counts > 20/hpf

Studies Evaluating Medications in Irritable Bowel Syndrome

Medication	Intervention	Control intervention	Outcome measured	Results	Efficacy
Amitriptyline [56]	Prospective, double-blind, randomized, placebo-controlled parallel-group study ×4 weeks treatment following 1 week observational period Amitriptyline ($n=43$) 10 mg/day <35 kg, 20 mg/d >35 kg ×4 weeks in patients with **irritable bowel syndrome**, functional abdominal pain, or functional dyspepsia	Placebo ($n=40$)	**Overall response to treatment** (self-assessment of pain relief and sense of improvement) **Effect on psychosocial traits** **Effect on daily functioning**	**Overall response to treatment** Self-reported feeling better: Amitriptyline: 63 % Placebo: 57.5 %	Not significant ($p=0.63$)
Tricyclic antidepressant (TCA) [11]	Retrospective review of patients prescribed amitriptyline or imipramine ($n=146$): for **irritable bowel syndrome**, dyspepsia, functional disorder of the intestine	N/A	**Overall response to treatment**	**Overall response to treatment** Responded: 78.6 % for average of 10.73 months Amitriptyline Responded well: $n=40$ (IBS $n=27$) Did not respond: $n=8$ (IBS $n=5$) Stopped due to adverse effects: $n=3$ (IBS $n=3$) Imipramine Responded well: $n=41$ (IBS $n=15$) Did not respond: $n=5$ (IBS $n=4$) Stopped due to adverse effects: $n=2$ (IBS $n=1$)	TCAs effective in treating functional gastrointestinal disorders in pediatric patients per author report; no statistical analysis

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Medication	Intervention	Control intervention	Outcome measured	Results	Efficacy
Amitriptyline [63]	Prospective, randomized, double-blind, placebo-controlled study **Weeks 0–2** Complete data question packets **Weeks 3–10** Amitriptyline ($n=16$) 30–50 kg: 10 mg at bedtime 50–80 kg: 20 mg at bedtime 80 kg: 30 mg at bedtime **Weeks 11–13** No medication, complete data question packets	Placebo ($n=17$)	**Symptom checklist** **Pain rating scale** **Visual analog scale** **IBS quality of life scale** completed at baseline and at 2, 6,10, and 13 weeks	**IBS quality of life scale** Patients taking amitriptyline significant more likely to have improvement in quality of life score at 6, 10, and 13 weeks	6 weeks: $p=0.019$ 10 weeks: $p=0.004$ 13 weeks: $p=0.013$
				Symptom checklist Patients taking amitriptyline more likely to experience reduction in IBS-associated diarrhea at 6 and 10 weeks	6 weeks: $p=0.029$ 10 weeks: $p=0.029$
Lactobacillus rhamnosus GG [57, 70, 71]	Meta-analysis evaluating utility of *lactobacillus GG* in functional disorders (all three articles enrolled patients with **IBS**, $n=20$) 3×10^9–10×10^{10} CFU twice daily for 4–8 weeks	Placebo	**Change in abdominal pain severity and/or frequency score**	**No pain or decrease in pain intensity** Treatment compared to placebo in patients with functional abdominal pain ($n=290$) RR 1.31 95 % CI 1.08–1.59 NNT 7 95 % CI 4–22	Significantly higher rate of responders; favors treatment
				No pain or decrease in pain intensity Treatment compared to placebo in patients with IBS ($n=167$) RR 1.70 95 % CI 1.27–2.27 NNT 4 95 % CI 3–8	Favors treatment
				Frequency of pain Frequency reduced in treatment compared to placebo in patients with IBS ($n=117$) SMD −1.04 95 % CI −1.43 to −0.65	Favors treatment
				Perception of pain intensity Pain intensity decreased in treatment compared to placebo in patients with IBS ($n=117$) SMD-0.60 95 % CI-0.97–0.23	Favors treatment

Symbiflor® (SF2) (*Escherichia coli* bacteria) [72]	Observational study of pediatric patients given SF2 until symptom improvement or up to 3 months ($n=203$) Children: 1 × 10 drops/day diluted in water taken midday Adolescents: Dose can be increased to up to 30 drops/day	None	**Frequency of pain**	**Frequency of pain** Frequent 4–11 years: Pre- $n=66$ to post- $n=38$ Occasional 4–11 years: Pre- $n=7$ to post- $n=38$ Frequent 12–18 years: Pre- $n=63$ to post- $n=33$ Occasional 4–11 years: Pre- $n=6$ to post- $n=28$	Statistically significant
			Stool frequency/day	**Stool frequency/day** 4–11 years: Pre- 1.69 to post- 1.11 12–18 years: Pre- 1.56 to post- 1.00	Statistically significant
				All other IBS-associated symptoms (bloating, passage of gas, etc.)	Statistically significant
Peppermint oil [74]	Prospective, randomized, double-blind study × 2 weeks ($n=21$) >45 kg: 374 mg three times daily 30–45 kg: 187 mg three times daily	Placebo ($n=21$)	**Change in symptom severity scale**	**Improvement in symptom severity scale** Peppermint: 76 % Placebo: 19 %	Significant improvement ($p<0.001$)
			Mean severity of pain symptoms	**Improvement in severity of pain** Lower in peppermint group	Significantly lower in severity ($p<0.03$)
Rifaximin [10]	Prospective, double-blind, placebo-controlled study of patients with functional dyspepsia ($n=11$), **IBS** ($n=41$), functional abdominal pain ($n=23$), or abdominal migraine ($n=0$) that were given rifaximin ($n=49$) Rifaximin: 550 mg orally three times daily × 10 days	Placebo ($n=26$)	**Symptom improvement (overall and individual symptoms)** **Hydrogen levels** (not reported here) **Methane levels** (not reported here)	**Symptom improvement** Specifics not mentioned in study	All values not significant between groups

Studies Evaluating Medications in Rumination

Medication	Intervention	Control intervention	Outcome measured	Results	Efficacy
Anxiolytics or antidepressants [81]	Medications given not reported ($n=6$) Medications combined with nutritional support, biofeedback, relaxation techniques, cognitive therapy ($n=12$)	None	Not reported	**Symptom resolution or improvement** 10/12 patients	Combination treatment effective
Baclofen [82]	Case series (only reported in abstract by third party author) ($n=10$)	None	**Regurgitation symptoms**	**Regurgitation symptoms** Decrease in symptoms by 50 %	N/A
			Rumination episodes	**Rumination episodes** Decrease in rumination episodes by 65 %	

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Music Therapy in Pediatrics: Clinical Indications for the Treatment of Functional Symptoms

27

Clare H. Arezina

Abstract

Music therapy is the clinical, evidence-based use of music interventions to accomplish individualized goals within a therapeutic relationship. Cross-culturally, music has been acknowledged as an integral part of healing rituals throughout recorded history. Music therapy is distinct from music listening and other types of "music medicine" due to its emphasis on individual assessment and treatment planning, goal setting, and the importance of a therapeutic relationship through implementation by a trained clinician. In pediatric settings, music therapy goals are aligned with the goals of child- and family-centered medical treatment: music therapists utilize elements of music and rhythm in interventions to support pediatric patients and families in successfully coping with stressors, particularly stressors inherent to illness and hospitalization. Successful coping includes the amelioration of functional symptoms that arise in association with anxiety, agitation, poor arousal/emotional regulation, low mood, refusal to cooperate/comply with treatment, and pain. This chapter provides a brief review of the literature supporting the efficacy of music therapy in pediatric medical settings, as well as a case example and suggested guidelines for referral.

Keywords

Music therapy • Pediatric • Coping • Child development • Complementary • Supportive care

Throughout recorded history, music has been acknowledged as an integral part of healing rituals in cultures around the world: for example, traditional Eastern medicine incorporates music and sound as an essential aspect of healing. "Medicine men" in a number of preliterate cultures use chanting, drumming, and/or dance prominently in healing rituals [1]. A Biblical passage credits David's music for the healing of King Saul [1, 2]. In the USA, accounts of the use of music in medicine date back to the late 1700s, when case studies were published touting the

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amelioration of fever and depression with music [3]. By the late 1800s, Blackwell Psychiatric Hospital conducted more extensive studies, documenting the use of live music played by professional musicians to calm its patients [3].

As defined by the American Music Therapy Association, music therapy is the clinical and evidence-based use of music interventions to accomplish individualized goals within a therapeutic relationship. Music therapy should be provided by a credentialed professional who has completed an approved music therapy program [4]. Music therapy was formally established as an allied health profession after World War II, largely due to the experience of staff at Topeka Veterans' Hospital. When volunteer musicians played in the wards, physicians and nursing staff noted positive changes in patients' mood, arousal, and ability to interact with others. Academic training programs for music therapists were established shortly thereafter, and clinical research in the field is ongoing [3, 5, 6]. Today, music therapists are specially trained to conduct a comprehensive individual needs assessment and devise and carry out a music-based treatment plan that considers individual strengths, preferences, and needs. Using clinically researched techniques, music therapists utilize the rhythmic, melodic, harmonic, and social-emotional principles of music to support and encourage changes in mood and behavior. Physical, cognitive, social, and emotional goals may be addressed through music therapy interventions with a wide variety of populations, from premature infants to the elderly. A successful music therapy intervention results in improvements in both functional outcomes and quality of life [6–8].

With recent advances in brain imaging and understanding of neurochemical processes, evidence is still emerging about the mechanisms by which music listening leads to pleasure, arousal, increased immunity, and positive social affiliation [9]. Clinical research in music therapy has shown that music therapy interventions can positively impact heart rate, respiratory rate, blood pressure, oxygen saturation levels, motor rehabilitation, perception of pain and anxiety, and feelings of well-being in both children and adults [8, 10].

Reduction/normalization of heart rate and respiratory rate are linked to the body's tendency to *entrain* a variety of responses with rhythms in the environment; *entrainment* refers to a change in rhythm in consideration of another rhythm. Even at the autonomic level, the body can match its rhythms (heart rate, respiration rate, motor movements) with rhythm presented externally [11, 12]. Given this entrainment of responses (coordination of a rhythm in the body with rhythm in the environment), it is possible to decrease/normalize heart rate and respiration rate via a steady, gradual increase or decrease of the tempo of music presented. For example, a music therapist might be asked to see a patient whose heart rate is too rapid. The music therapist would begin by introducing a music stimulus at a fast tempo to coordinate with the patient's elevated heart rate (for a young child, perhaps Twinkle Twinkle Little Star accompanied by a guitar). Over the next several minutes, the music therapist would gradually slow the tempo a few beats per minute to encourage an entrainment response of the patient's autonomic nervous system. Such a gradual shift in the type of musical stimulus that produces a change in autonomic or behavioral response has been defined as the *iso principle*. Similarly, arousal and emotional response can be shaped through the iso principle [3, 10, 11, 13–15]. Although there is limited research in this area, there is some evidence to suggest that agitation (motoric evidence of overarousal) can be mitigated by music [16, 17]. As a music therapist working in a pediatric setting, this writer has clinically observed a number of patients whose agitation was reduced through the use of entrainment and the iso principle.

Children in medical settings experience a variety of physiological, medical, developmental, and psychosocial stressors, all of which can negatively impact their treatment course [18–21]. As has been addressed extensively in the medical literature and elsewhere in this text, medical settings are inherently distressing to children: an unfamiliar environment, personnel, and language and a loss of familiar routines and loss of control lead to increased fear and anxiety, which both are informed by and impact experience of pain and other functional symptoms (i.e., difficulty breathing, agitation,

anxiety, low mood, etc.). A variety of pharmacological and non-pharmacological interventions can be employed to address children's functional symptoms of distress and increase their ability to cope with stressors; music therapy is one such non-pharmacological intervention [6, 8, 19–25].

For most children, listening to music is an inherently pleasurable experience; even young infants show preference for culturally familiar music [26, 27]. Music is an integral part of most entertainment media for children (music/sound toys, songs on children's television programs, etc.), and most educational experiences for children include group music, either formally or informally [27, 28]. Participation in live music, through either physically active participation or passive listening, is an inherently multisensory experience. Music therapists can structure interventions to provide an appropriate variety of sensory input with respect to patients' level of arousal and ability to integrate sensory information (which may be impacted by the physiological stress of illness and hospitalization) [24, 29–31]. In addition to providing a variety of auditory stimuli through the use of familiar and unfamiliar songs, musical styles, and instruments, music therapy interventions can provide tactile stimulation (vibration of air/instruments, patient manipulating instruments, therapist utilizing/facilitating caregiver touch as part of songs), visual stimulation (watching the therapist play instruments or sing, visually attending to instruments played by self or others), proprioceptive stimulation (manipulation of instruments, participation in finger-plays or dancing), and vestibular stimulation (movement to music, including self-directed dancing or adult-directed rocking). In providing an integrated multisensory experience, music therapy can minimize perception of other physiological stimuli, including pain and anxiety, and may thus lead to positive interoception, or a general feeling of well-being [32].

In addition to distraction through multisensory engagement, music therapists can structure sessions that offer patients meaningful choices, invite them to engage in novel or previously enjoyed pleasurable experiences, provide a safe medium for verbal or nonverbal self-expression, actively engage children at their cognitive-developmental level, and provide new learning opportunities as appropriate—all of which can positively impact patients' hospitalization experience and ameliorate distress [29–31, 33–38]. Careful consideration of a child's developmental level is essential in the provision of any intervention to address functional symptoms, as stressors inherent to the medical setting differentially impact children according to their physical, emotional, and cognitive development [19, 20, 25]. Music therapy interventions can be uniquely structured by a trained professional to address individual needs accordingly (see Table 27.1 for examples of common concerns related to hospitalization and music therapy interventions to address each of them).

Notably, there are a number of music therapy applications to decrease stress response and increase coping behaviors for infants in the Neonatal Intensive Care Unit (NICU) as young as 32 weeks gestational age (GA) [39, 40]. There has been significant research to support the use of music therapy to encourage sensory regulation, limit overstimulation, and encourage positive responses to stressors in these patients with underdeveloped sensory processing and self-regulation systems [39–43]. Although applications in the NICU are similar to those with typically developing infants, there are a number of important considerations in working with premature infants, most significantly their difficulty processing/tolerating sensory stimulation. Music therapists who work in the NICU setting should have advanced training in evidence-based applications specifically for that population, which include intensive training in recognizing stress responses in neonates, responding to stress responses appropriately, and providing effective interventions that appropriately engage the neonates' ability to tolerate multisensory stimulation while respecting their low threshold for overstimulation [39, 40].

Practical Application

While there is some research evidence to support the use of prerecorded music and the provision of music by minimally trained performers

Table 27.1 Music therapy interventions to address developmental concerns during hospitalization

Chronological age	Developmental stage [18]		Developmental concerns of hospitalization [18]	Music therapy interventions [14–17, 21, 25, 29–31, 36–38]
	(Erikson)	(Piaget)		
Infant Birth–18 months	Trust vs. Mistrust	Sensorimotor	• Loss/mistrust of caregiver relationship	• Music play, singing, movement to music between patients and caregivers facilitated by therapist • Use of family-preferred prerecorded music to establish routine (bath, bedtime)
			• Sensory experience (physical "hurts")	• Live music to provide a positive multisensory experience – Singing – Rocking – Instrument play (emerging cause and effect learning)
Early childhood 2–3 years	Autonomy vs. Shame and Doubt	Preoperational	• Lack of opportunities for mastery	• Offer clear structure/routine in session (greeting, closing, cleanup) • Sing familiar, preferred songs—allow children to fill in words as they know them • Provide developmentally appropriate instruments to provide rewarding "success" – Child-directed motivation/ incentive for participating in treatment – "Music breaks" for sips/bites of food – Wind instruments to encourage deep breathing/expiration, reduce oral aversion
			• Loss of control	• Offer meaningful choices verbally/nonverbally (What song? Which instrument? What color? How many? Quiet or loud?) • Basic instrumental improvisation (child-directed start/stop play)
Preschool 3–5 years	Initiative vs. Guilt		• Magical thinking/ misattribution	• Expressive play – Musical improvisation – Instrument exploration – Guided songwriting – Music-facilitated dramatic play – Redirection/reframing of concerns verbally or musically
School age 6–11 years	Industry vs. Inferiority	Concrete Operational	• Lack of opportunity for achievement	• Provide challenging, rewarding musical opportunities – Simple/complex rhythm imitation – Beginning music skill development – Therapist- or patient-directed instrumental improvisation – Songwriting – Product creation (music video/album) – Music-facilitated relaxation training using imagery, rhythm, deep breathing
Adolescence 12–18 years	Identity vs. Role Confusion	Formal Operational	• Loss of identity; new identity as "sick person"	• Offer opportunity for identity formation as "musician" – Instrument instruction (guitar, piano, etc.) – Songwriting/improvisation – Product creation (music video/album) • Lyric analysis to engage higher level thinking, relation of personal issues to those of others

(sometimes referred to as "music medicine," "music listening," or "therapeutic music" but sometimes erroneously labeled "music therapy" in the research literature), there are well-documented contraindications for the global provision of music in any setting. Especially in vulnerable populations, there is concern for overstimulation and unintended negative emotional response [6, 7, 16, 17, 39, 40]. Additionally, music therapy interventions have consistently demonstrated larger effect sizes than other types of music medicine [8]. Music therapists are specifically trained in multimodal assessment and development and maintenance of effective therapeutic relationships and are thus able to anticipate, prevent, and/or mitigate negative outcomes by altering or stopping the music/intervention and addressing concerns as they arise in the moment [7].

In the USA, music therapists are nationally board certified; training includes a minimum of a bachelor's degree in music therapy (which includes extensive course work in musicianship, human development across the lifespan, and basic psychology, neurology, physiology, and music perception, in addition to training in appropriate music therapy assessment, design, intervention, and evaluation of outcomes), at least 1,200 h of directly supervised clinical work, and successful performance on a board exam [44]. Training programs around the world follow a similar model; in many countries, music therapists are trained at the master's level, similarly to psychotherapists.

Appropriate referrals for music therapy may include pediatric patients with any of the following characteristics:

- Anxiety and functional symptoms that can develop in association with anxiety
- Agitation/hyperactivity
- Poor eye contact/interaction with staff members
- Pain not well managed pharmacologically
- Low mood
- Extended hospitalization (>3 days)
- Acute neurologic event (brain surgery, head trauma)
- Developmental delay/sensory processing disorder
- Concern for developmental regression/stagnation
- Many stressful/traumatic events during hospitalization (multiple attempts for intravenous line placement and/or phlebotomy, diagnostic tests, etc.)
- Lack of response to other available psychosocial interventions (Child Life, play therapy, etc.)

Case Study

Anna, a 5-year-old girl, was hospitalized with severe viral pneumonia. Anna had been a generally healthy, typically developing child prior to this extended hospitalization; she was initially admitted to an outside non-pediatric facility for more than a week and then sent to the Children's Hospital when her condition was not improving. The music therapist met Anna a few days into her hospitalization at the Children's Hospital upon referral from the Child Life Specialist. The referral noted that since her arrival, Anna required multiple "pokes" for IV starts/attempts, blood draws (both of which had also been done multiple times at the outside facility), and recent placement of a chest tube and that she was unable to leave her room due to isolation precautions. In addition, it had been noted by her physicians that Anna seemed to develop increased shortness of breath whenever she was told that she is about to undergo a medical procedure. The Child Life Specialist reported that the patient was in very poor spirits, yelling at hospital staff and her parents, and had very vocally rejected all attempts by Child Life to provide preferred toys and activities at bedside. Anna was also refusing to complete breathing exercises (incentive spirometry), even with a behavioral incentive plan in place (sticker chart/prizes).

When the music therapist met the patient, Anna was initially resistant to the therapist's presence but showed visual interest in instruments and asked if just the instruments could stay. With permission from Anna's parents, who were both at bedside, the therapist offered the patient opportunities to improvise musically on

instruments, varying the level of the therapist's musical involvement in her play as Anna allowed. As Anna played with a drum, she verbally identified it as an "angry" sound and spontaneously stated that she was angry because the nurses like to poke her and hurt her. Her nurse was in the room to check her IV medications, and the therapist asked him if he likes to poke kids—he stated that he does not. The music therapist reinforced for Anna that nurses do not like to give pokes but that sometimes pokes have to happen to help kids get better. Anna seemed to accept this answer, continuing to play the drum, sometimes accepting the therapist's play on other instruments, and sometimes choosing to exert control to prevent the therapist from playing with her (either by "growling" or stopping her own play).

The therapist then introduced a variety of wind instruments—kazoo, harmonica, and slide whistle, demonstrating each—Anna stated that she "hated" the kazoo and the harmonica was "too loud" but smiled when she heard the slide whistle. Anna was eager to have her name written on the whistle and listened as the therapist told a story about how parents know when slide whistles are "too tired" and have to "go to bed" on a shelf in the room; Anna quickly asked her parents where the slide whistle could sleep at home. Anna readily played with the music therapist, taking turns in a call-and-response exercise, e.g., making eye contact and waiting for her turn while the music therapist played. When it was her turn, Anna was able to play similarly to the therapist as well as create her own sounds for the therapist to imitate. She was able to breathe deeply and extend the amount of time she could sustain a sound on the whistle within the same session. She willingly helped to clean up and said goodbye without prompting when the session was over.

Over the course of the 30-min session, the patient's affect had changed dramatically. At the beginning of the session, Anna was withdrawn and growled/shouted at the therapist but smiled broadly and showed age-appropriate interaction by session's end. Anna was discharged a few days later—her parents and nurses noted a positive change in her mood, no further episodes of shortness of breath in association with medical procedures, and willingness to play the slide whistle (and complete her breathing exercises!) throughout the rest of her hospital stay.

Questions

1. Which is the best explanation for Anna's perception that the nurses "like to poke her"?
   (a) Anna saw a nurse smile during a blood draw.
   (b) Anna is in the preoperational stage of cognitive development.
   (c) Anna is trying to make sense of her experience.
   (d) Anna overheard a nurse say that the nurse likes to poke kids.
   (e) b and c.
2. Which of the following may account for Anna's willingness to talk with the music therapist?
   (a) Physically engaging in music play was pleasant to Anna.
   (b) Having control over music-making lessened Anna's anxiety.
   (c) Characterizing a sound was less personal than talking about her own feelings.
   (d) Anna felt comfortable with the music therapist.
   (e) All of the above.
3. What was the benefit of a music therapist working with Anna, rather than the Child Life Specialist providing musical toys?
   (a) The Child Life Specialist was too busy to spend 30 min with Anna.
   (b) The music therapist is specially trained to engage and respond to Anna through music.
   (c) Anna did not like the Child Life Specialist.
   (d) Anna might have broken/misused musical toys without proper supervision.
   (e) Anna might have ignored the musical toys.

Answers

1. (e): At 5 years of age, Anna is in the preoperational stage of cognitive development—magical thinking and misattribution occur at this age when children begin to develop basic reasoning skills but are not able to consider all possibilities. In trying to make sense of her experience, Anna misattributes the nurses' willingness to poke her to a desire to hurt her.

It makes sense to her that people do not do things they do not like to do: if a nurse is willing to hurt her, he or she must like it.

2. (e): Anna likely responded to the music therapy intervention because she felt comfortable in the session: the music therapist allowed Anna to be in control of the music, provided a familiar, pleasurable experience, and gave Anna an opportunity to express herself nonverbally. (Nonverbal expression may be similarly achieved through other expressive therapies such as play therapy or art therapy.) For most children, emotions are first expressed nonverbally, through play: Anna's music was angry, and the therapist validated her anger in listening to her music and responding to it. Anna was willing to talk about her feelings with the therapist because the therapist had already validated her musically.
3. (b): Anna may have enjoyed playing with musical toys in the absence of a trained music therapist, but it was only through the musical interaction with the therapist that Anna was able to express her feelings, develop a trusting relationship with a member of the hospital staff, and have her misconceptions about her experience addressed.

Conclusions

Music therapy is a non-pharmacological intervention that can successfully address a variety of functional symptoms in pediatric patients through the goal-directed use of music- and rhythm-based interventions within a therapeutic relationship. Trained music therapists are uniquely qualified to assess individual needs and design and implement interventions that utilize the rhythmic, melodic, harmonic, and social-emotional principles of music to affect both mood and functional outcomes in patients. There is a growing research base to support the use of music therapy to impact a variety of positive health outcomes, including heart rate, respiration rate, blood pressure, oxygen saturation levels, motor rehabilitation, perception of pain and anxiety, and feelings of well-being in both children and adults.

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Play-Family Therapy: A Biobehavioral Team Approach to Chronic Medical Symptoms

28

Dottie Higgins-Klein

Abstract

When there is concern that a child's chronic medical problems involve emotional issues, the literature supports an approach based on a biobehavioral model that includes the family in the treatment. A therapy process that attends to both the individual psychology of the child and the underlying dynamics within the family system usually results in improvement or resolution of problems that do not respond well to medical treatment alone. This chapter provides a brief summary of how play therapy has evolved to integrate family system issues and presents a case study demonstrating the value of Mindfulness-Based Play-Family Therapy.

Keywords

Biobehavioral • Family systems • Family therapy • Mindfulness • Mindfulness-Based Play-Family Therapy • Play therapy • Sandtray therapy

While medical care providers are able to offer a variety of interventions that help children recover and maintain health, they also see that children sometimes need more than physical healing, and there is a growing awareness that children's somatic problems may be associated with emotional and relational issues. Somatic problems that may be triggered by emotional distress include chronic stomach pains, migraine headaches, tics, asthma attacks, and sleep disturbances. Such symptoms are often associated with psychological conditions, such as anxiety, depression, or chronic anger. There is a range of experiential triggers that can give rise to these emotional states in children, including unresolved early trauma, loss from the death of a loved one, abuse, bullying, the impact of foster care, and stories and images in the media. Such emotional states may be exacerbated by family dynamic issues such as early destructive parenting, the child's loyalty split between parents in conflict, or a child's feelings of powerlessness.

A biobehavioral family model takes the viewpoint that a child's psychosomatic situation is encompassed within the family structure: therefore, it acknowledges the influence of family members' reactivity patterns on each individual's

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physiological, emotional, and behavioral responses to emotional stimuli [1]. The effect of whole-family emotional health on individual physiology is apparent in the observed linkage between family-focused interventions and improvement in somatic symptoms, for example, in work with children's asthma [2].

In order to address the emotionally based aspects of health issues, pediatricians, family practice physicians, and nurse practitioners can refer children for play therapy that involves both the individual child and his family. In the therapeutic framework of Mindfulness-Based Play-Family Therapy (MBPFT), the combination of play therapy with family therapy offers optimal healing for the child, as well as for the whole family. Through the play therapy component, the child can gain healing of current emotional challenges and of the effects of past developmental deficits. Whereas adults are developmentally capable of reworking emotional problems through talk therapy, the child's mind does not yet have this capacity—for him, the metaphors of play allow more access to his psyche than is available through words alone. The family therapy component of the work leads to the repair of conditions in the family system that are major factors contributing to the child's problems.

A Brief History of Play Therapy

The field of play therapy has progressed from its original tendency to see the child's problems in isolation to a point of view that includes a family systems perspective, which is based on how relational reverberations may be at the root of physical and emotional health. Credentialed play therapists are master's- or doctoral-level clinical psychotherapists that are required a minimum of 150 h of play therapy training and supervision, as well as continuing education. They are generally licensed, for example, as marriage and family therapists, social workers, professional counselors, or psychologists.

Early pioneers of play therapy, in the 1930s and 1940s, Melanie Klein [3] and Anna Freud [4] used play primarily to help the child talk about his problems. They observed the child and then made interpretations based on the play.

Client-Centered Play Therapy, introduced by Virginia Axline in 1947, emphasizes the role of the therapist as following the child's lead in the play [5]. She based this well-documented play therapy modality on the reflective listening methods of Carl Rogers [6]. Thus, children play freely and do not have to defend against the therapist's interpretations. Garry Landreth made a major contribution by refining the theory and application of the client-centered approach throughout his career [7].

Filial Play Therapy, developed in the 1960s by Bernard and Louise Guerney, amplified the growing connection between play therapy and the family system by teaching parents to hold weekly play sessions at home, thus adopting a client-centered approach toward their children [8].

Theraplay, presented in 1971 by Ann Jernberg, also involves parents directly in their children's therapy [9, 10]. Theraplay is a particularly valuable method for working with children who have attachment issues or who have autism spectrum disorder complicated by significant impairment of communication.

Floortime, also known as DIR—Developmental, Individual Differences, Relationship-based Model—was developed by Stanley Greenspan and published in 1992 [11]. This modality allows parents to work directly with their children in their play therapy. It is particularly valuable for children with developmental delays, including those associated with autism spectrum disorder. It is often combined with sensory integration therapy provided by occupational therapists. In an example of healthy integration of medical and psychological fields, occupational therapists receive training at our center in MBPFT to develop competence in addressing the emotional issues that children exhibit.

Family Play Therapy, introduced in 1994 by Eliana Gil [12] and Charles Schaefer and Lois Carey [13], made a major leap in connecting the worlds of Family Therapy and Play Therapy. The authors encouraged play therapists to involve the parents more in treatment and urged family therapists to be more creative and playful when

working with children. When working with parents and their children, the therapist leads the activities and addresses issues in family dynamics through engagement with what happens in the session. Lenore Terr set a solid theoretical framework for using play therapy for trauma [14], and Eliana Gil has spent much of her career advancing the work with trauma, including sexual abuse [15].

With their book in 2001 [16], Deborah Killough McGuire and Donald McGuire offered a theoretical framework that encouraged play therapists to include parents more systematically in the treatment of the child. Their work increased awareness of the environment of the child's daily life and advanced the natural progression of the field toward parent education. In recent years, the Association for Play Therapy has initiated a strong campaign to provide empirical data in support of the efficacy of play therapy [17]. In 2005, Bratton, Ray, and Rhine presented an analysis of 93 controlled studies from a 47-year-period ending in 2000, which supports the inclusion of parents in the child's therapeutic process, concluding that "using parents in play therapy produced the largest effects." Their analysis also indicates the broad efficacy of play therapy: "Play Therapy appeared equally effective across age, gender and presenting issue" [18].

Mindfulness-Based Play-Family Therapy (MBPFT) [19] has furthered the therapeutic correlation, in both its theory and its interventions, between the child and his family system. One practical example of this is its recommendation that the therapist meet with a parent and the child for 20 min preceding every play session, in order to discuss whatever is important in the lives of the child and his family. This empowers the parent to develop skills to talk about and address the challenges of life. Another example is the inclusion of family members in the initial evaluation sessions. There are also regular, at least monthly, mindful parenting meetings, without the child present, in which parents are invited to reflect on their own contribution to the problems that are bringing their family to therapy.

The play therapy component of an MBPFT session offers a time of direct engagement between therapist and child through the medium of the child's pretend play. Child-led play therapy allows the child to go to a profoundly deep place in his psyche through his play metaphors. In a way similar to mindfulness meditation, it offers access to a state of security even in the face of inner turmoil. This secure state can bring healing of the feelings, including those of powerlessness, that are intimately connected with a history of trauma. This combination of a family systems approach with play therapy offers clinicians an excellent option for the treatment of the emotional aspects of children's health issues.

The following case study demonstrates how MBPFT utilizes parental and sibling involvement and how the child may heal during individual play therapy. This case shows how the therapist works to address the root level of a child's individual and relationship-based emotional problems, and how that leads to an alleviation of physical symptoms. This study has been drawn from several actual cases and the names have been changed.

Case Study

A message on the answering machine requested that I return a call to Dr. Rao. He is a pediatrician who refers children for MBPFT when he observes that their chronic medical problems seem to be rooted in emotional or family-related disturbances. I was surprised because the message concerned Carlos, a former client whom Dr. Rao had referred to me 3 years earlier, when Carlos was 5 years old. The doctor had received a desperate call from Carlos's mother, calling on the strong suggestion of the third grade teacher, Ms. Smith, who complained that Carlos's oppositional behaviors at school had escalated following winter break. He had been disturbing other children on the playground, intimidating one child whom he clearly did not like, and disobeying teachers when they directed him to perform routine tasks. The teacher had tolerated these erratic behaviors for a couple of weeks and was now strongly suggesting to his mother that his doctor should prescribe Ritalin. Dr. Rao wanted my opinion and requested that I see the family. I was grateful that he called me before prescribing medication, since

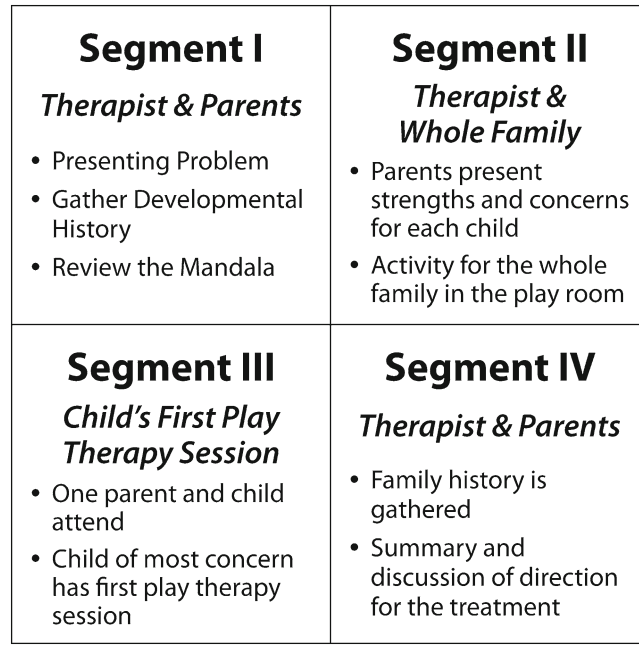

Fig. 28.1 Four-segment evaluation in mindfulness-based play-family therapy (Copyright © 2013 Dottie Higgins-Klein, Family and Play Therapy Center, Philadelphia, USA)

my memory of this child was that he did not display symptoms of attention-deficit/hyperactivity disorder. I wondered how things had changed this much for him.

The next message on my machine was from Ana, Carlos's mother. She was quite anxious and described his aggressive, out-of-control behaviors, as well as his recurrence of stomach aches. When Carlos had first come to MBPFT 3 years earlier, he had just started attending a new school. He was having stomach pains, unwilling to separate from his mother, and refusing to ride on the school bus. His parents were cooperative with the therapy, and within 6 months, Carlos had adjusted to riding the bus, separating from his mother, and participating in his new classroom.

Although we had completed the Four-Segment Evaluation 3 years earlier, I decided to repeat some of it, in order to learn about the developments that had occurred during the intervening time (Fig. 28.1). It informed me about what was happening, not only with Carlos but also with his parents and his younger sister, Sophie. This process is undertaken as a collaboration rather than as an intrusive examination of the past and present. My routine is to have the first evaluation session with the parents without the child, so they are free to tell me anything without the child hearing it. Ana and her husband, Ernesto, reviewed the previous notes concerning Carlos's early developmental history. His history did not bring up issues of concern from his first 3 years. He was delivered vaginally without medical complications. His developmental milestones were within the normal range: he walked by his first birthday, had tantrums at two, and was toilet trained by 3 years. There was no history of early trauma.

However, when we turned to more recent developments, I learned that Carlos's maternal grandparents had both died in a car accident just 6 months ago. This was a major loss for the entire family. Ana was feeling depressed. Carlos himself had been close to his grandparents. I offered my condolences and sat with presence as Ana quietly cried, while Ernesto put his arm around her. Ana was then able to continue, and we completed the final section of the Developmental and Social History Form (19, see form in Appendix). Parents rated a list of present concerns about their now 8-year-old son:

- Issues of mild concern included temper tantrums and low self-esteem.
- Issues of moderate concern included aggression and conflict with schoolmates, overeating (he was somewhat overweight), disobedience, mood changes, and frequent displays of frustration.
- Issues of serious concern included anxiety and fears. His parents noted that although they did see some of his fears and anxiety at home, these problems were occurring more at school than at home.

Finally, I reviewed the Mandala (Fig. 28.2), particularly noting the stages of play therapy and mentioning that things might get worse before they get better.

At our second evaluation session, I introduced the family to a collection of miniature toys, organized neatly on five shelves, in categories that represented all aspects of life, such as people, animals, nature, scary objects, and symbols of spiritual experience. This material provides a way to introduce the entire family to an experience of play therapy. These four family members were invited to select three miniature toys each.

They worked as a group and placed their spontaneous choices into a rectangular tabletop sandtray to create a sand picture, and then they took turns to make up a pretend story. In the sand story, they told about a monster that lived in a cave. At night, it came out to scare the townspeople, who were depicted by a pig family of four, selected by Sophie. The monster, a two-headed dinosaur chosen by Carlos, was invisible to the parents—only the children were able to see it. They all enjoyed creating an elaborate story that contained themes of fear, aggression, death, and humor. Taking the last turn, Carlos ended the story with the mother and father pigs yelling at the baby pigs for sword fighting and rolling in the mud (Fig. 28.3).

A sand story is an essential part of the evaluation process. It provides a simple structure in which the therapist can share an experience with the whole family and can learn about family dynamics and personalities. I observed that mother and son still had a close alignment, as they had in the past. I saw a strong connection between father and daughter. While the children were getting along fairly well with each other, I also noticed a typical amount of sibling rivalry. Since MBPFT cares about everyone, I noted that I would need to encourage Sophie to find more of a voice for her own concerns.

Carlos and his mother came to the third evaluation session together. This session had two parts: first, the three of us met together in Talk Time; then, Carlos had a play therapy time. Talk Time began with a brief check-in about what was going well and what the child and parents had enjoyed about their week. This was followed by the discussion of one problem or concern.

Ana began the Talk Time by sharing that they had enjoyed a cousin's birthday party during the weekend. Carlos chimed in with his agreement. Then she told me that, as was usual on Sunday evening, Carlos had had severe stomach cramps and did not want to go to school the next day. After some discussion, I slowed the process and invited Carlos to do a visualization to address his physical symptoms, saying, "Close your eyes and feel into your stomach area. How does it feel now?" He described it as a big knot that "breathes like an eyeball. It is red and gets bigger and smaller." After allowing a half minute for both of us to feel this sensation, I asked whether the eyeball wanted to say anything to him. Carlos said, "No." I asked, "If the eyeball had a voice and it was very smart and could tell you why you don't want to go to school, what would it say?" To this he answered, "Oh, it would say that I don't have friends at school." I responded, reflecting his words, "Hmm… you don't have friends at school…" Carlos opened his eyes and said that his two best friends from last year were moved to a different class, he had not made any friends this year, and some kids teased him at lunchtime. This made him mad, so he teased other kids, in ways that annoyed them, though he wished he could just play with them.

For the play therapy part of this evaluation session, Carlos's mother went to the waiting room. After he explored toys in half of the playroom, Carlos chose to make a sand picture. He selected several different sports figures and arranged them in various groupings in the dry sandbox.

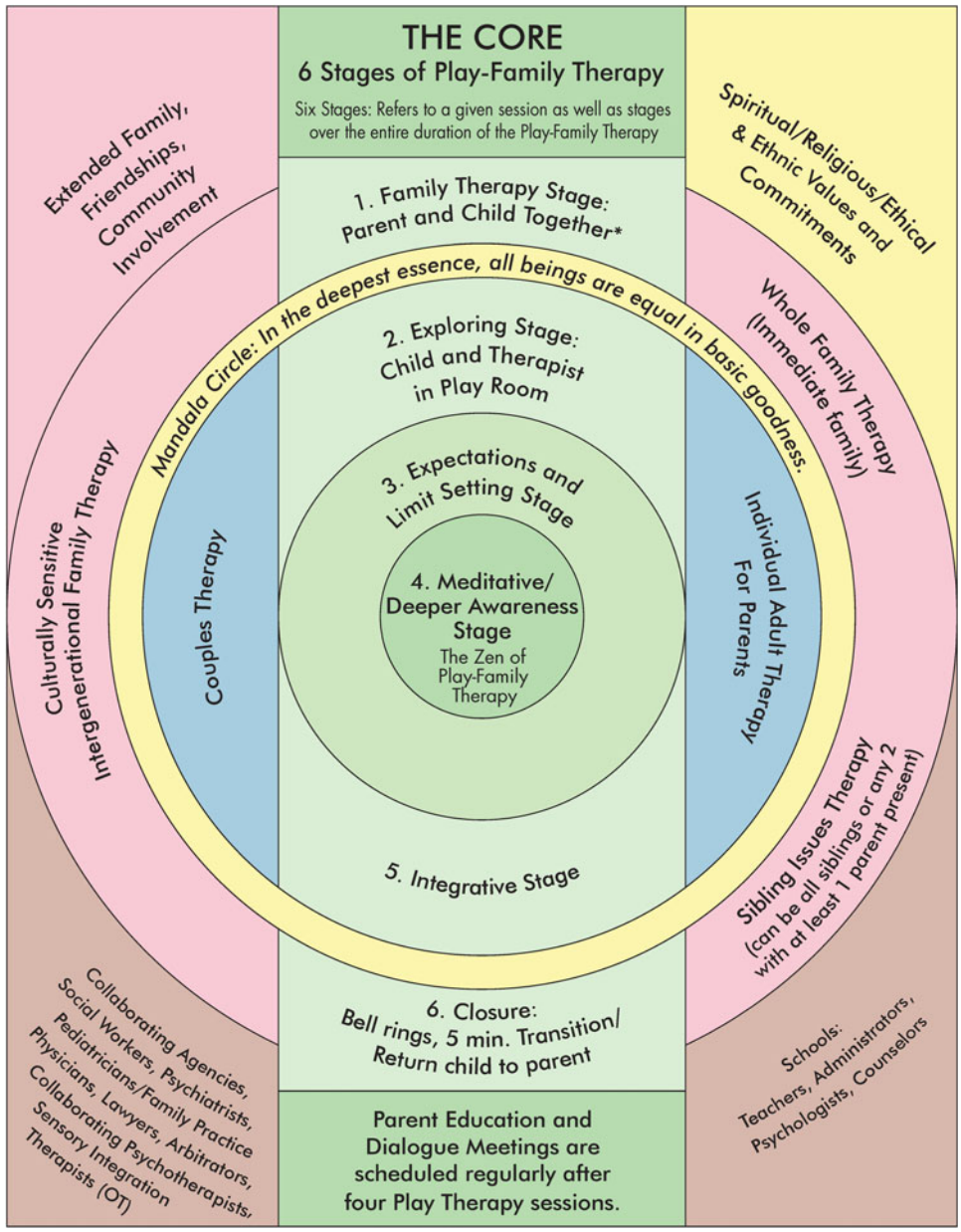

Fig. 28.2 Mindfulness-based play-family therapy Mandala (Copyright © 2013 Dottie Higgins-Klein, Family and Play Therapy Center, Philadelphia, USA)

There were clusters of sports figures playing basketball, soccer, and football, and a boy on a skateboard. Behind a fence in the lower left corner, there was a lone soccer player kicking a huge soccer ball with his back to the other players (Fig. 28.4). Carlos did not want to say anything about this sand picture. I let him know that it would be fine to use words or not—he could decide. In future sessions, as he engaged more in the pretend of the play therapy, he began to describe his anxieties and fears through the metaphor of his stories. Over time, this process not only helped him to formulate and express his feelings, but it also allowed him to face into them and to heal.

Fig. 28.3 Second evaluation session: family sand story

Fig. 28.4 Third evaluation session: Carlos's first Sandtray

At the fourth evaluation session, I normally ask each parent about their family histories and discuss a recommended course of action. In this case, we reviewed and updated Ana and Ernesto's middle-class, second-generation Puerto Rican family histories. While a particular family constellation is discussed here, the MBPFT model is adaptable to the many ethnicities that the clinician may encounter in practice. Ethnicity and Family Therapy is a book that serves as an invaluable clinicians' resource for multicultural awareness [20].

I again offered my compassion for their bereavement. What I learned of significance was that since the maternal grandparents' deaths, the couple had grown apart. They had developed a tendency toward hostile arguments with one another, sometimes occurring in front of the children. I paid attention to the reverberations of stress that exacerbated their children's emotional issues. When asked, the parents admitted that Carlos's symptoms increased when they themselves felt more stressed. I acknowledged that this was a valuable observation for them to make.

With the four evaluation sessions completed, we began the course of Play-Family Therapy. Carlos's parents took turns bringing him to the weekly sessions. Prior to each of Carlos's play therapy sessions, we held the usual 20-min Talk Time. During this time, we addressed in detail one concern from among Carlos's emotional, behavioral, physical, or relational issues. Once a month, there was a Mindful Parenting Meeting, attended only by the parents. In addition to my providing feedback about the themes of the play therapy, these sessions offered an opportunity to help the parents recognize ways in which their own emotional state had become a formative component in the emotional climate of the whole family. Sessions focused both on the child's behaviors and on healthier ways to engage in family life.

After about 2 months, Ana and Ernesto reported success in keeping their arguments private. We had a family session that addressed the children's fears that their parents could divorce. The parents reassured them that this was not at all their intent. Carlos's problems at school decreased significantly. He stopped behaving aggressively toward other children and began to listen to his teacher's directives. I had explained to the parents that this is what we call symptom relief and that by continuing the play-family therapy we would be promoting a healing deeper and more lasting than just the relief of these symptoms. During the fourth month of play therapy, Carlos began to descend more into the Deeper Awareness Stage. His play quietly enacted the pain of feeling trapped. He used a scary man who was stuck inside a wheel-like cage and variations of that metaphor (Fig. 28.5). I quietly witnessed these intense, pretend play themes over a half-dozen sessions. His feelings of frustration and hopelessness gradually changed until he felt more empowered, and the man emerged from his circular cage. In the same sense, so did Carlos. At that point, he was moving toward more lasting change and entering the integration stage of play therapy.

Parallel to Carlos's work on his feelings of helplessness, he began to speak up more honestly at home. This sort of opening behavior is associated with the Integration Stage. It can sometimes be disconcerting to parents when a child does this, but it demonstrates that he is not afraid to be truthful about his life pain. Such honest dialogue helps build trust in the parent–child relationship. Defining "respect" in this Hispanic family led to interesting cultural discussions. The parents were able to see that when Carlos was afraid to speak up at home, he was repressing his true feelings and acting them out at school. On the other hand, the raw, blaming anger that Carlos was beginning to express was not in line with his parents' idea of respect. We had some additional family meetings, at which each member described his or her style of displaying anger. We worked on respectful ways for everyone to express anger. Both children began to feel safe enough to directly address their father. They told him that when he gets angry, they are afraid and hide together in their bedroom. Hearing his children voice this directly helped motivate Ernesto to work on how to better manage his anger. It was equally important that Ernesto see that I did not intend to condone Carlos's speaking to him without respect. Respect shown by a child to his father was a significant part of his cultural value system, and, as the therapist, I was sensitive to giving it attention as a resource.

Fig. 28.5 Deeper awareness stage: scary man stuck inside a wheel-like cage

What Happened with This Family?

In MBPFT, the play is kept in a pretend context. The child speaks of family problems only when parents join him at Talk Time. This helps the child avoid feeling disloyal to parents and helps the parents learn valuable skills in dealing with hard-to-talk-about subjects with their child—a skill that continues after the family ends therapy. The metaphoric quality of the play therapy time allowed the playroom to be a venue for Carlos to feel his fears, sadness, and confusion. Carlos's main play themes expressed his wish for inclusion with friends, concern about being teased for his weight, feelings of anger and blame toward his father, and a fear that his parents might divorce. These metaphoric themes correlated with the actualities that were discussed during Talk Time.

Reflections of the Treatment and Long-Term Follow-Up Provided for Carlos and His Family

- I kept contact with the teacher and encouraged the parents to take an active role in addressing concerns about Carlos's socializing at school. Carlos needed teacher intervention to help him join in free-play activities.
- A few brief conversations with Dr. Rao proved to be valuable for our teamwork. Families that may hesitate to seek outside help are often very responsive to their physician's advice. Rather than prescribing medication, Dr. Rao encouraged the parents to pursue the play-family therapy. After 3 months, Carlos's somatic symptoms were significantly reduced, and it became clear that he did not need medication for either hyperactivity or anxiety.
- The parents made a commitment to continuing therapy for Carlos beyond the initial symptom relief because they understood that deeper healing required more time. The early stage of therapy builds a basis of trust that often creates a temporary calm. As seen in this case, such trust allowed the emergence of buried feelings that Carlos had been afraid to express at home. His parents learned that it is better to have the parent–child conflict addressed directly in the home rather than have him displace his feelings by misbehaving at school. The experience was that things became worse before they got better. If therapy is stopped during "the quiet before the storm," the chance for deeper healing may be arrested.
- The Talk Time meetings helped Carlos develop skills for impulse control and for using words instead of disturbing behavior to assert his feelings with other children. We also addressed the stage of normal fears that happens with this age child. At 8 years of age, a child understands more about living, and it is normal for him to go through a fearful stage.
- Strategies were developed to help Carlos engage friends both at school and at home.
- Dr. Rao and I both recommended that the whole family have a few meetings with the nutritionist in his medical practice. Since there was a link between Carlos's emotional distress and his misuse of food, a constructive attention to nutrition could contribute to the creation of a healthy emotional state.
- The parents recognized that their recently acquired habit of responding to stress by showing anger toward one another in front of the children could be contributing to the development and persistence of Carlos's destructive behaviors. Their admission that they sometimes avoided speaking to one another for a few days at a time led to their acknowledgment that both children were sensitive to their displays of anger, both overt and covert. At occasional meetings with the whole family, Sophie was able to verbalize that she, too, was worried about her parents. The meetings helped me confirm that there was indeed split loyalty, with the children taking their mother's side against their father. This had been partly caused by Ana's demeaning comments about Ernesto, made in front of the children. "Split loyalty is one of the most challenging areas of therapy work, and one of the most common. It is useful for parents to know the degree of harm that can be caused by split loyalty—that it is at the root of self-destructive behavior. Constant exposure to split loyalty is one reason that a child's behaviors can

become very intense and uncontrolled" [19]. It is essential to address this family therapy issue in order to achieve deep healing.

- The couple discussed how the sudden deaths of Ana's parents had affected their relationship. Both began to work on their own anger management. After Carlos finished his therapy, Ana requested some individual therapy. She used the sandtray modality for adults [19] to help her with her grieving process. The adult method of sandtray can include metaphoric stories but relies more heavily on communication of the reality of what is happening as it works with unconscious material. The therapist listens to what the sand picture means to the client, avoiding interpretation. Ana was surprised at how comforting and profound this healing method could be.
- Carlos's anxiety was substantially reduced, as were his somatic symptoms during this 9-month treatment period. Two years later, he was behaving quite well at school. He now willingly played on a community soccer team that he had resisted joining prior to the play-family therapy. During the following year, Carlos did not need to see Dr. Rao except for his annual check-up. At that time, the doctor noticed that Carlos's overall confidence and self-esteem had greatly improved.

Questions

1. The Four-Segment Evaluation offers the following benefits EXCEPT:
   (a) The therapist becomes informed about current problems.
   (b) The therapist can experience the family dynamics at the second session.
   (c) The child of most concern can experience the play therapy and begin to express his thoughts and feelings through metaphor.
   (d) The pediatrician attends the first meeting.
   (e) Parents may begin to better understand how the child's struggle with anxiety reflects their own anxieties.
2. Some of the symptoms that may be addressed through Play-Family therapy are:
   (a) Anxiety
   (b) Sadness
   (c) Angry behaviors
   (d) None of the above
   (e) All of the above
3. All of the following are true EXCEPT:
   (a) Multi-team collaboration of the family with the pediatrician, play-family therapist, teacher, and nutritionist offers optimal healing potential.
   (b) There are many different kinds of play therapy, with varying degrees of parental involvement.
   (c) Play Therapy and Sandtray Therapy for children are completely different modalities.
   (d) MBPFT addresses all the family members, even though one child may be seen in individual play therapy.
   (e) Early on in treatment, a child's symptoms can be helped through play therapy; however, continued play-family therapy offers a more lasting level of healing.
4. The following are ways that a pediatrician or family practice physician can be helpful:
   (a) By encouraging a family evaluation with a play-family therapist when there are symptoms rooted in emotional issues
   (b) By listening to the concerns of teachers who notice changes in a child's behavior
   (c) By collaborating with the play-family therapist before deciding on medication for behavioral or emotional symptoms
   (d) All of the above
   (e) None of the above

Answers

1. (d): Communication between the therapist and the pediatrician generally involves only one or two brief phone calls. There is no need for the pediatrician to be present at the family meetings.
2. (e): Play-Family Therapy offers options for addressing a wide range of emotional issues. It respectfully attends to a child's symptoms and involves understanding of the wider family issues so that root-level healing can occur.
3. (c): Play Therapy is considered a solid theoretical framework for helping children address

emotional problems and behavioral issues with the help of a caring therapist. Sandtray Therapy for children is a subcategory of play therapy using specific trays of sand and hundreds of miniature objects to create pictures and stories. It relies on the theoretical framework of play therapy. Sandtray for teens and adults assumes that the therapist has a grounding in reliable psychotherapeutic theories. Sandtray is not an independent theoretical construct.

4. (d): Family members highly respect the recommendations of their pediatricians. When there are chronic physical symptoms that may be rooted in emotional or family dynamic issues, families are more likely to seek help if their physician encourages a referral for play-family therapy.

Conclusions

Some physical symptoms in a child may be rooted in psychological causes. Trauma, loss, developmental delays, and difficult attachment experiences, among other things, all can create deep psychic issues that may manifest through somatic and behavioral symptoms. In such cases, physical treatment and immediate behavioral correction, while perhaps necessary, do not, by themselves, reach far enough into the child's psyche to yield reliable, long-term healing. The ideal approach is a combination of medical care with systems thinking, as exemplified by MBPFT, which includes cooperative engagement with the extended family and the child's school.

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Appendix

Chapter 19: Cognitive-Behavioral Therapy for Youth with Functional Somatic and Internalizing Symptoms

Physicians seeking additional training in CBT may access webinars, podcasts, videos, and therapist manuals sponsored by the following associations: American Psychological Association (http://www.apa.org), Association for Behavioral and Cognitive Therapies (http://www.abct.org), and Anxiety and Depression Association of America (http://www.adaa.org). These organizations also hold annual conventions that include live demonstrations of therapy techniques and research presentations on the latest application of CBT techniques to individuals with functional somatic symptoms. In addition to the resources offered by these organizations, we strongly suggest forming partnerships with psychologists or social workers practicing CBT within academic medical settings who can offer ongoing supervision and consultation. This additional support will be helpful when implementing more complex techniques, like exposure, and when dealing with challenging cases or family situations. Forming partnerships with CBT specialists will also provide a convenient and trusted resource for patients that may benefit from an external treatment referral.

Handout for families to help determine whether a therapist specializes in CBT:

Questions for Finding a CBT Practitioner

- *What is your theoretical orientation?* Therapists who answer cognitive, cognitive-behavioral, behavioral, or evidence based will be best equipped to provide state-of-the-art CBT. Answers such as "eclectic" or "integrative" or "psychodynamic" suggest that the therapist is not trained in CBT.
- *Do you do exposure therapy?* Exposure is another term for "facing your fears," during which patients practice anxiety-provoking or previously avoided situations. This also often includes practicing realistic thinking to help youth engage in sometimes difficult exposure exercises.
- *Do you give homework exercises?* CBT therapists routinely give patients homework or practice exercises in between sessions.
- *Are parents involved in treatment?* While parents are often less involved with treatment of adolescents, they should always generally know what their child is expected to be working on in order to provide additional coaching and support the child's efforts.
- *Do you give the child advice about how to handle difficult situations?* You want to hear that the therapist provides active suggestions about what to do and that they will practice

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these techniques together during the session. You will want to avoid practitioners who say they prefer to listen, support, or "be a sounding board" for the child, and do not try to push the child beyond their comfort zone.

Chapter 20: The Use of Biofeedback and Neurofeedback in Pediatric Care

Healthcare professionals who would like to learn more about the application of biofeedback, or who would like to acquire entry level biofeedback skills, are encouraged to attend the meeting of one of the two primary biofeedback societies in the USA, the Association for Applied Psychophysiology and Biofeedback (AAPB), and the International Society for Neurofeedback and Research (ISNR).

AAPB provides both pre-conference workshops and a scientific meeting on both general biofeedback and neurofeedback (http://www.aapb.org). Invited guests present state-of-the-art reports on relevant basic science ranging from cardiovascular rehabilitation to neuroimaging. The conference includes an exhibit hall where both equipment manufacturers and instrument vendors staff booths and provide attendees with demonstrating of current instrumentation systems. The vendors are eager to allow attendees to try out instrumentation, in order to facilitate the selection of the optimal biofeedback system. AAPB has three publications. The journal *Applied Psychophysiology and Biofeedback* is the leading peer reviewed and indexed scholarly publication in the field, and *Biofeedback: A Clinical Journal* provides journalistic style articles describing many of the latest developments in both biofeedback and neurofeedback. The publication *Neuroconnections* is a joint publication of AAPB's neurofeedback division with the ISNR and highlights advances in neurofeedback and quantitative EEG assessment.

ISNR also provides a combination of workshops and scientific presentations, with an emphasis on neurofeedback, but a growing inclusion of general biofeedback topics as well (http://www.isnr.org). The ISNR conference also features an exhibit hall, and many of the current developers of neurofeedback systems and software are present to discuss the state of the art in neurofeedback and quantitative EEG instrumentation. ISNR publishes the *Journal of Neurotherapy*, which reports both clinical and scientific advances, and co-sponsors the *Neuroconnections* publication.

Both professional societies are "user friendly," with leading researchers and practitioners open to interacting with new attendees and students. Both organizations have Foundations developed to provide students with travel scholarships to attend the meetings and to support doctoral research in psychophysiology, biofeedback, and neurofeedback.

Newcomers to the field of biofeedback and neurofeedback should also be aware of the Biofeedback Certification International Alliance (BCIA), which provides certification in biofeedback and neurofeedback, in the USA and abroad (http://www.bcia.org). The BCIA has published "blueprints of knowledge," comprehensive lists of skills and knowledge that any entry-level practitioner should master, for general biofeedback, neurofeedback, pelvic floor biofeedback, and heart rate variability biofeedback. BCIA provides certification for individuals in general biofeedback, neurofeedback, and pelvic floor biofeedback. It also provides a BCIA "Certificate in Heart Rate Variability Biofeedback," for individuals who document attendance in workshops covering the HRV blueprint.

BCIA, AAPB, and ISNR collaborate to promote an emphasis on minimal professional standards for practice, as well as "best practices." All three organizations provide a variety of resources for education and training, including publications, online bookstores, banks of research articles, webinars, and CE credit for reading journal articles.

There are several sources of comprehensive training workshops in biofeedback and neurofeedback, which are designed to prepare individuals both for professional practice and the BCIA certification exams. The STENS corporation provides 1-day introductions to biofeedback and neurofeedback, and comprehensive training programs for general biofeedback (5 days), heart

rate variability (2 days), and neurofeedback (4 days) (http://www.STENS.com). Stress Therapy Solutions offers 4-day neurofeedback certification workshops, as well as specialized workshops on quantitative EEG, "Z-score-" based neurofeedback training, and advanced clinical neurofeedback (stresstherapysolutions.com). Saybrook University's School of Mind-Body Medicine offers a Certificate program in biofeedback and neurofeedback (http://www.saybrook.edu/mbm), for Saybrook students and for non-matriculated health professionals. Alliant International University, East Carolina State University, Sonoma State University, Truman State University, and Widener University (among others), all offer BCIA certified training programs. The BCIA Website provides a regularly updated list of University and commercial biofeedback and neurofeedback training programs that have qualified as covering the BCIA blueprints.

Chapter 21: Hypnosis for Treatment of Functional Symptoms in Children

Clinical hypnosis is best learned through active participation in hypnosis workshops sponsored or endorsed by medical hypnosis organizations. Hypnosis instruction is appropriate and encouraged for various staff members who might be employed at a primary care practice or at pediatric specialty centers including physicians, fellows, residents, physician's assistants, nurse practitioners, master's level nurses, psychologists, respiratory therapists, physical therapists, dietitians, child life specialists, and master's level social workers.

Basic-level workshops typically are taught over 3–4 days and provide at least 20 h of training, including at least 6 h of supervised skill development in small group hypnosis practice sessions of workshop participants. Following such an initial introductory course, the healthcare provider can be ready to start employing hypnosis successfully with patients who have common diagnoses that tend to respond well to hypnosis, i.e., anxiety, headaches, irritable bowel syndrome, and vocal cord dysfunction. Follow-up intermediate- and advanced-level hypnosis courses often are helpful.

Training for clinicians caring for children and adults is offered several times a year around the USA by the American Society of Clinical Hypnosis.
Website: http://www.asch.net
Telephone number: (630) 980-4740

Training for clinicians caring for children and adults is offered annually in the USA by the Society for Clinical and Experimental Hypnosis.
Website: http://www.sceh.us
Telephone number: (617) 469-1981

Training for clinicians caring for children is offered annually in Minnesota, USA, by the National Pediatric Hypnosis Training Institute.
Website: http://www.nphti.org
Telephone number: (800) 776-8636

Training for clinicians caring for children and adults is offered every 3 years around the world by the International Society of Hypnosis.
Website: http://www.ish-hypnosis.org
Telephone number: +39 06 854 8205

Training for clinicians caring for children and adults is offered several times a year throughout Europe by the European Society of Hypnosis.
Website: http://www.esh-hypnosis.eu
Telephone number: +44 114 248 8917

Training for clinicians caring for children and adults is offered several times a year throughout Australia by the Australian Society of Hypnosis.
Website: http://www.ozhypnosis.com.au
Telephone number: +61 2 9747 4691

Chapter 22: Guided Imagery for Functional Disorders

The Academy of Guided Imagery may be a valuable resource for learning more about the use of imagery:

Academy for Guided Imagery
P.O. Box 2070
Mill Valley, CA 94942
800-726-2070
http://www.acadgi.com

Chapter 23: Acupuncture for the Treatment of Functional Disorders in Children

There are multiple avenues for further training in medical acupuncture for physicians within North America (Please see below). Many courses offer a portion of the curriculum as home study, so that physicians may continue to practice while pursuing training in medical acupuncture. In the USA, requirements for practice of medical acupuncture by physicians are variable amongst individual states. The authors recommend a course in medical acupuncture geared toward the training of physicians that incorporates at least 300 credit hours. The majority of training programs available in the USA are geared to meet this minimum training period. Additionally, physicians in the USA can attain a Board certification (DAMBA) in medical acupuncture administered by the American Board of Medical Acupuncture (ABMA) requiring a written examination after completion of their training and a minimum practice period.

Licensed acupuncturists are required to attend a minimum of 3–4 years of training in order to obtain a Diplomate of acupuncture, while the designation of a licensed acupuncturist (LAc) is awarded by a state regulatory board after minimal criteria have been met by an applicant. Passing of a certification examination administered by the National Certification Commission for Acupuncture and Oriental Medicine (NCCAOM) is a requirement for non-physician acupuncturists' licensure to practice Oriental Medicine in a majority (43) of states across the USA.

The following is a list of American Board of Medical Acupuncture (ABMA)-endorsed training programs for physicians considering a training and board certification in medical acupuncture:

Acupuncture Integrative Medicine Program
Tri-State College of Acupuncture
80 Eighth Avenue, Suite 400
New York, NY 10011
(212) 242-2255
http://www.tsca.edu/site/prospective/doctors/
Director: Mark D. Seem, Ph.D., Lac

The Art and Science of Acupuncture: Basic and Advanced
University of Miami
PO Box 016960 (D-79)
Miami, FL 33101
(305) 243-4751
http://www.cam.med.miami.edu
Director: Janet Konefal, Ph.D.
Complete Course in Medical Acupuncture for Physicians
Inst of Continuing Medical Education
PO Box 70169
San Juan, Puerto Rico 00936
(787) 751-5979
http://www.colegiomedicopr.org
Director: Luis E. Vazquez Zayas, M.D.
Contemporary Medical Acupuncture for Health Professionals
McMaster University, Office of CME
1280 Main Street West, Room #HSC-2U
Hamilton, ON L8S 4K1
(905) 521-2100 Ext.75175
http://www.acupuncturecourses.com
Director: Alejandro Elorriaga Claraco, M.D.
Administrative Coordinator: Valerie Cannon
Medical Acupuncture at Downstate
SUNY Downstate Medical Center
450 Clarkson Avenue, Box 1244
Brooklyn, NY 11203
(718) 270-4563
http://www.downstate.edu/ocme
Director: Tsai C. Chao, M.D.
Medical Acupuncture for Physicians
Helms Medical Institute
2520 Milvia Street
Berkeley, CA 94704
(510) 649-8488
http://www.hmieducation.com
Director: Joseph Helms, M.D., F.A.A.M.A.
Medical Acupuncture Program: An Evidence Based Approach to TCM
Michael G deGroote School of Medicine
1200 Main Street West, Room MDCL-3112
Hamilton, ON L8N 3Z5
(905) 546-5500
http://www.acupunctureprogram.com
Director: Ken Trinh, M.D.
Program Administrator: Marsha Walker
Medical Acupuncture-Level I and II
Acupuncture Foundation Canada Institute
2131 Lawrence Avenue East #204
Scarborough, ON M1R 5G4
(416) 752-3988
http://www.afcinstitute.com
Director: Cheryll A. Kwok

Structural Acupuncture for Physicians
Harvard Medical School
Department of Continuing Education
Boston, MA
(781) 391-7518 Ext. 240
http://cme.hms.harvard.edu/index.asp?SECTION=CLASSES&ID=00302317&SO=N
Director: Joseph F. Audette, M.D.
Course Administrator: Kathy Marino

Chapter 24. Mindfulness Meditation for Children

Further information about mindfulness meditation for children is available:

Training Opportunities

Dialectical Behavioral Therapy (DBT): http://www.behavioraltech.org

Acceptance and Commitment Therapy (ACT): http://contextualpsychology.org/act

Mindfulness-based Stress Reduction (MBSR): http://www.umasssmed.edu/cfm/, http://cme.ucsd.edu/mindfulness/index.html

Mindfulness-based Relapse Prevention (MBRP): http://depts.washington.edu/abrc/mbrp/

Resources

Books

Biegel GM. The Stress Reduction Workbook for Teens. New Harbinger Publications, CA. 2009

Fontana D and Slack I. Teaching meditation to children: A practical guide to the use and benefits of meditation. Watking Publishing, 2007

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Lantieri L. and Goleman D. Building Emotional Intelligence: Techniques to Cultivate Inner Strength in Children. Sounds True, Inc., 2008

Vallely SW. Sensational Meditation for Children. Satya International. 2008

Weierbach J and Phillips-Hershey E. Mind Over Basketball: Coach Yourself to Handle Stress. Magination Press. American Psychological Association, 2008

CDs

Gina Biegel. Stressed Teens: Mindfulness for teens – Meditation Practices to Reduce Stress and Promote Well-Being. 2009

Florence Meleo-Meyer. Meditations for Children.

Florence Meleo-Meyer. Cool Minds.

Amy Salzman. Still, Quiet Place: Mindfulness for Young Children, 2004

Chapter 25: Adapting Yoga for Children and Adolescents with Functional Disorders

Many resources regarding yoga are available:

Yoga Alliance (YA), the accrediting body for yoga teachers lists minimum training standards for teachers and schools: https://www.yogaalliance.org/

Yoga Alliance (YA), minimum training standards for teaching yoga to children: http://www.yogaalliance.org/ya/c/Standards/Childrens_Yoga_Standards.aspx

International Association of Yoga Therapists (IAYT), the professional association for yoga therapists and schools lists minimum training standards for yoga therapy. Also provides links to yoga research articles and IAYT publications: http://www.iayt.org/

National Center for Complementary and Alternative Medicine (NCCAM), yoga information page: http://nccam.nih.gov/health/yoga/introduction.htm?nav=gsa

Child focused yoga teacher training programs and interventions programs:

Yoga Ed: http://www.yogaed.com/, http://www.yogaed.com/resources.html

Yoga for the Special Child: http://www.special-yoga.org/

Birthlight: http://www.birthlight.com/

Teen Yoga: http://www.teenyoga.co.uk/

The Wellness Initiative: http://www.wellnessinitiative.org/

Yoga as a Complementary Therapy for Children and Adolescents: A Guide for Clinicians: http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2945853/

Books for Teaching Yoga to Children

1. The Yoga Adventure for Children, Helen Purperhart, Hunter House Inc., Alameda, CA, 2007. (Ages 4-12).
2. Kid Yoga: Fun with a Twist, Juliet Pegrum, Sterling Publishing Inc., New York, NY, 2004.
3. Yoga for Teens: How to Improve your Fitness, Confidence, Appearance and Health – and Have Fun Doing It, Thia Luby, Clear Light Publishers, Sante Fe, NM, 2000.
4. Fly Like a Butterfly: Yoga for Children, Shakta Kaur Khalsa, Sterling Publishing Inc., New York, NY, 1950. (Kundalini Yoga)
5. Storytime Yoga: Teaching Yoga to Children Through Story, Sydney Solis, The Mythic Yoga Studio, Boulder, CO, 2006.
6. My Daddy is a Pretzel: Yoga for Parents and Kids, Baron Baptiste, Barefoot Books, Cambridge, MA, 2004.
7. Babar's Yoga for Elephants, Laurent De Brunhoff, Harry N. Abrams, Inc., New York, NY, 2002.
8. Twist: Yoga Poems, Janet S. Wong, Simon & Schuster Children's Division, New York, NY, 2007.

Chapter 27: Music Therapy in Pediatrics: Clinical Indications for the Treatment of Functional Symptoms

For information about music therapy training, please consult:

American Music Therapy Association: http://www.musictherapy.org

World Federation of Music Therapy: http://www.musictherapyworld.net/WFMT/Home.html

Chapter 28: Play-Family Therapy: A Biobehavioral Team Approach to Chronic Medical Symptoms

For more information about play therapy, the following web sites may be consulted:

http://www.AAMFT.org: American Association for Play Therapy—Systems trained family therapist—some are trained in play therapy as well. Site offers a registry for family therapists.

http://www.A4pt.org: Association for Play Therapy offers credentials and a registry for play

http://www.cacpt.com: Canada Association for Play Therapy

http://www.Fptcenter.com: Family & Play Therapy Center offers Mindfulness-Based Play-Family Therapy Training throughout the world with our interactive, live on line, state-ofthe-art technology (iLOC). We refer world-wide. On site is in Philadelphia and includes a counseling center.

http://www.iaptp.ie: Ireland Association for Play Therapy

http://www.israplaythrpy.org: Israeli Play Therapy Association

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