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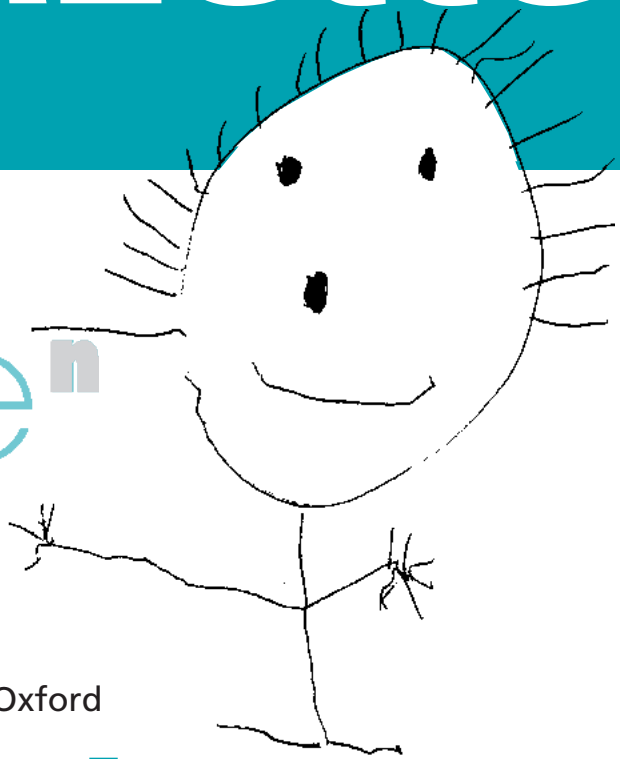

Paul L. Harris, Oxford

Feeling Happy, Feeling Sad

How children talk about and hide emotion

It is possible to study emotion without paying much heed to our everyday capacity for identifying and talking about emotion. Psychologists can focus on the way that people betray their emotions through facial expressions, through various telltale patterns of behavior, or through psychophysiological reactions. Indeed, whether we look back at Freudian theory or at contemporary neuropsychological approaches to emotion, there is a line of argument which says that our lay understanding and awareness of emotion is limited, and certainly a narrow window through which to study emotion scientifically. Nevertheless, adults are far from being oblivious to emotion. They are often painfully aware of their own emotions and those of other people. In this article, I describe the way that children first start to develop such awareness. In particular, I focus on two different aspects of that development: children's growing ability to talk about emotions and their appreciation of the fact that what people actually feel may differ from the emotion that they overtly express.

Talking about Emotion

A major index of children's self-awareness is their ability to put emotions and feelings into words. Recent studies of language development have charted the emergence of this ability. Henry Wellman and his colleagues [1] studied the spontaneous remarks of 2-, 3- and 4-year-olds recorded in the course of their everyday interaction with parents and siblings. They examined all those utterances in which children referred to an emotion, whether experienced by themselves or oth-

er people. This analysis revealed that 2-year-olds talk systematically about a small set of emotional states, both positive (feeling happy or good, laughing, and feeling love or loving) and negative (feeling angry or mad, feeling frightened, scared or afraid, and feeling sad or crying). They talk most often about their own feelings, but also mention those of other people and those of dolls, stuffed animals and made-up characters. Thus, almost as soon as they are able to talk, children supplement the nonverbal communication of emotion through language – they put their own feelings and those of other people into words.

A common assumption about early emotion utterances is that they should not be regarded as *reports* but rather as *expressions* of feeling, on a par with expressive exclamations such as "Wow" or "Ouch" (Wittgenstein [2]). On this assumption, one would expect children's early use of emotion terms to be mainly concerned with their current emotions. After all, if talk about emotion is primarily aimed at expressing felt emotion, there should be few references to emotions that are not being experienced at the time of the utterance, i.e. emotions that belong in the past or might occur in the future. However, even at 2 years of age, only about half of children's emotion utterances are concerned with present feelings. Their other utterances are concerned with past, future, and recurrent feelings, and this pattern continues among 3- and 4-year-olds. Indeed, the Wittgensteinian approach appears to be inaccurate not just for the expression of emotion but also for the expression of pain: even in this context, children talk about noncurrent feelings – pains that they have already experienced or might experience. All this suggests that we can reasonably think of children's emotion utterances as genuine reports; they are not a kind of verbal grimacing or smiling. In line with this conclusion, close analysis of children's emotion utterances shows that they can be mainly categorized as descriptive or discursive statements; they are sometimes used with a pragmatic end in view – to seek help or comfort, or to change another person's emotional state – but that is not their primary function.

Nevertheless, one might still wonder if such young children are capable, not just of offering an emotional report, but of offering a report that is accurate. To answer this question, Richard Fabes and his colleagues [3] watched preschool children as they engaged in free play at their day care center. The observers remained as unobtrusive as possible until they spotted a child overtly expressing an emotion, whether it was happiness, anger, sadness or distress. At that point, the observers would approach another child standing close by, one who had witnessed the expression of emotion but had not actually caused it, and ask what had happened. The reports supplied by children in response to these requests proved to be relatively accurate. For example, even 3-year-olds – the youngest age group questioned – provided reports of what the target child was feeling and what had provoked the reaction, which were in agreement with the observers' own observations about two-thirds of the time. Five-year-olds were even more accurate: their reports agreed with those of observers more than three-quarters of the time. In sum, we may conclude that young children talk about emotion, they often do so in a descriptive rather than an expressive or manipulative fashion, and their reports are fairly accurate. We can now look more closely at sources of variation within this general pattern.

Talkative Parents, Talkative Children

Consider a child whose father frequently talks about emotion, using it as a way to draw out the child's own feelings, to comfort the child, or to call the child's attention to the emotional consequences of his or her actions for other members of the family. Consider, on the other hand, a father who is relatively silent about the child's own feelings, and does not use language to highlight the emotional repercussions of the child's actions. It would not be surprising if the first child were more disposed to talk about emotion than the second, and indeed be more attuned to the relatively subjective nature of emotion – the fact that people feel differently about the same event. Research exploring this issue has begun to uncover an intriguing pattern. First, in line with intuition, families do differ dramatically in the frequency with which they talk about emotion. For example, working with families from a wide socioeconomic range living in and around the city of Cambridge in England, Judy Dunn and her colleagues [4] observed that some children never mentioned feelings throughout an entire, hour-long period of observation, whereas others made more than 25 references to feelings in the same period; the range among the children's mothers was equally dramatic. Is there any indication that these variations in family discussion of emotion influence children's developing understanding of emotion? Dunn and her colleagues have found that the frequency with which preschool children engage in discussion of emotions and their causes is linked to their performance some months or years later on tests of psychological understanding. Thus, the frequency of such discussion predicts children's later ability to identify how someone feels – both over a relatively short interval, from 33 to 40 months and over a more extended interval, from 3 to 6 years.

How should we interpret this link? It is obviously possible that some children are naturally gifted as psychologists – they are alert to people's emotional states, seek out and participate in more conversations about emotion, and later on display a keen ability to imagine how other people feel and think, as tapped by standard tests of psychological understanding. However, it is also possible that family discussion of feelings nurtures the child's empathy and perspective-taking so that they are more sensitive to what other people might feel. One piece of evidence in favor of this idea is that children's perspective-taking is especially likely to be linked to family discussion that focuses on *why* someone feels a given emotion rather than on simply what they feel. How do such discussions about causation increase children's sensitivity to emotional expression?

One intriguing possibility is that such conversation provides children with a format for rerepresenting and organizing the episodes of everyday life, especially those that are emotionally charged. Armed with that coherent representation, children would then be better equipped to think through the psychological implications of the episode. We know for example that the period under scrutiny – 2 to 5 years – is one in which memories increasingly escape the veil that infantile amnesia imposes. Katherine Nelson [5] has argued that this is because children increasingly participate in conversations about past episodes, are thereby encouraged to rehearse and organize such episodes into a coher-

ent narrative, and eventually learn how to impose that structure on events that they experience even in the absence of a conversation partner. As Dunn [6] has pointed out, there are two pieces of evidence that fit in with this proposal. First, children often focus on emotionally charged events when they narrate what has happened to a family member. Second, there is good evidence that the coherent encoding of an event is important if children are to work out its psychological implications. Thus, Charlie Lewis [7] found that 3-year-olds who normally do poorly on a standard test of psychological understanding – the so-called false-belief task (see box) – perform much more accurately if they are asked to compose and recount the events triggering the false belief into a coherent narrative question, rather than listening passively to an adult narration of such events. In short, one intriguing possibility is that conversation teaches children how to organize everyday episodes, especially those that are emotionally charged, into coherent causally connected sequences. In turn, that mode of causal organization alerts children to antecedents and consequences of emotional reactions, so that children become more sensitive to what other people might be feeling.

A second possibility inheres, not in the narrative function of conversation, but in its wider epistemic implications. Much of our knowledge about other people's mental states arises in the course of conversation [8]. Conversation with other people teaches us that we know or believe something that our partner does not and vice versa. Indeed, conversation involves an ongoing acknowledgment of such variation in knowledge and belief. For example, realizing our ignorance, we ask for information from someone more knowledgeable. Realizing their ignorance, we explain, elaborate or inform, and so forth. This analysis highlights variation between conversation partners in their information base. But conversation goes beyond the exchange of information – it involves the sharing and comparing of attitudes, especially emotional attitudes. In discussing their experience with a conversation partner, children will be confronted by the fact that what they find frightening or humorous may evoke a very different reaction in a sibling or caretaker, and they may well be supplied with reasons for that divergence. On this hypothesis, conversation about emotionally charged episodes can highlight variation between partners in the way that they interpret the same situation, and thereby alert children to the subjectivity of emotion.

In future research, we may expect a more focused effort to examine these different hypotheses. In particular, it is important to find out whether it is the prompting of a causally coherent narrative structure and/or the opportunity to contrast and compare emotional attitudes within the framework of the conversational exchange that is associated with children's emotional understanding later on.

Hiding Emotion

Children learn not only to identify and talk about emotions, they also learn that emotions can be concealed, so that what someone expresses on their face may or may not correspond to what they really feel. Converging evidence shows that children become systematically aware of the potential distinction between actual and expressed emotion at around 5 years of age. To assess their understanding of this distinction, we can tell children simple stories involving a protagonist who encounters some upsetting obstacle or mishap (e.g. he trips over) but who also has a good reason to hide his distress (e.g. to avoid being called a crybaby by other children) and ask them about the emotional implications of such an encounter. In particular, we can ask what the story character will really feel, and what emotion he will express. Six-year-olds understand that the character might really feel upset but will try nonetheless to look happy or okay. They also understand that an onlooker is likely to be misled by this display, mistakenly thinking that the protagonist is not upset. By contrast, 4-year-olds are more likely to collapse the distinction between what the protagonist really feels and what he or she expresses. This age change is not just found among English-speaking children. Japanese and Indian children show a comparable pattern of development across this age period despite important differences among these countries in the cultural expectations that surround the display or concealment of emotion.

Does this mean that children will only start to hide their feelings at around 5 years of age? In fact, they show some tendency to hide their emotions before they fully understand the distinction between actual and expressed emotion. This has emerged in what has come to be known as the "disappointing gift" paradigm. Children are led to expect that they will receive a desirable gift or prize, but when they unwrap it, they discover that it is only a drab toy that they do not want. Their facial reactions to this disappointment are filmed. The key finding is that when children unwrap the gift alone, their disappointment is easily visible on their face. On the other hand, if they unwrap the gift in the presence of the adult who has given it to them, they express less overt disappointment. By implication, they try to hide their feelings. Nevertheless, if these same children are interviewed about the distinction between actual emotion and expressed emotion, they often cannot make the distinction.

Combining these various findings, it looks as if children start to conceal their emotions without being fully aware of what they are doing, certainly before they can conceptualize the distinction between what they feel privately and what they express to the outside world. Indeed, this shift from the practice of concealment to an understanding of its implications may be a necessary developmental sequence. Suppose that a child – without at first being fully aware of what she is doing – suppresses the outward expression of distress or disappointment. To the extent that she does this successfully, an adult is likely to be misled and wrongly identify the emotion as positive or at least neutral. Thus, the child feels upset or disappointed, and knows it; meantime, the responses of other people imply that she feels fine. At this point, the discrepancy between the child's actual experience and the emotion that other people attribute to her on the strength of her overt expression will be brought home to the child. It is interesting to note that this is especially likely if she is growing up in a family where talk about emotions is frequent. Extending the earlier line of speculation, therefore, we may anticipate that children who engage in frequent discussion of emotion will not only be better able to identify other people's emotions, they will also be more sensitive to the way in which the emotion that a person expresses may not match what that person really feels.

It is worth emphasizing the implications of the discovery that emotion can remain private. Children can feel resentful or victimized, and yet also know that those feelings may remain unknown to other members of their family. Indeed, in the complex, triangular relationships that sometimes arise within families, children may come to understand that one family member may express and talk about particular emotions with them, but conceal those same emotions from another member of the family, and insist that the child do likewise. In future research, as we begin to study children growing up in conflictual or dysfuntional families, it will be important to assess the ways in which these more complex patterns of acknowledgement and denial impinge on the children's developing awareness of emotion.

The false-belief task

In the false-belief task, children watch as a puppet puts some attractive item – for example some chocolate – into a container and then leaves. Next, children watch as a second puppet 'steals' the chocolate and moves it elsewhere. When the first puppet returns, children are asked where he will go to look for his chocolate. Most 4- and 5-year-olds, but few 3-year-olds, realize that he will at first – given his false belief about the location of the chocolate – search fruitlessly in the now empty box.

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Gerard J. Canny, Dublin

"health, and quiet breathing"[1]

How children live with asthma

Asthma is the most prevalent chronic disease of children, affecting 10–15% of the preschool population. Despite advances in understanding its pathogenesis and the development of improved medications, asthma remains a major source of morbidity among children, often because of underdiagnosis, undertreatment and insufficient knowledge among doctors and parents about the nature and possible treatments for the condition. Asthma is a common cause of hospitalization and emergency room visits, can significantly affect exercise tolerance, and may disrupt sleep. There is some evidence that the incidence of asthma is rising and that its severity may be increasing, especially in industrialized countries. Nevertheless, with close collaboration between physicians and parents, paying careful attention to the means by which the asthma can be alleviated and controlled, the vast majority of infants and young children should be able to lead active and full lives that are not limited by their disease.

What is Asthma?

There is no universally accepted definition of asthma. However, it is now generally considered to be a chronic inflammatory disease of the airways that leads to reversible airway obstruction and bronchial hyperreactivity to stimuli such as histamine and methacholine. From a clinical perspective, asthma should be suspected in any child with recurrent episodes of wheezing, coughing or dyspnea.

In the course of an allergic response, the local release of spasmogens and vasoactive substances (such as histamine and certain leukotrienes and prostaglandins) causes, in the airways, varying degrees of smooth muscle spasm, mucosal edema, mucus secretion and infiltration of eosinophils, T lymphocytes, mast cells and neutrophils. With the muscle contraction and airway narrowing, breathing becomes much more difficult, and there is often the characteristic wheezing, the high-pitched sound of air being forced through narrowed spaces. Other symptoms may include coughing and chest tightness.

Asthma is underdiagnosed. There seem to be several reasons for this: mislabeling (e.g. 'wheezy bronchitis'), a failure to appreciate the early onset of asthma in most children, and atypical symtomatology. A practical definition of asthma is therefore essential to optimize diagnosis in clinical practice. In a recent Canadian report [1] it was proposed that, "Any child, regardless of age, who presents with a history of recurrent episodes of wheezing, dyspnea, or chronic cough should be considered to be suffering from asthma until proven otherwise."

The Irritant Environment

A distinction should be made between *inducers*, i.e. factors which may cause asthma, and *triggers*, i.e. factors which may aggravate preexisting asthma.

Asthma seems to arise from complex interactions between genetic and environmental factors. The frequency of asthma is higher in children whose parents have asthma, and boys are twice as likely to develop asthma as girls, although this difference disappears by adolesence. Molecular genetic studies are beginning to identify genes that may be associated with atopy (an increased tendency to form IgE antibodies following exposure to environmental allergens), bronchial hyperresponsiveness and cytokine production. Recent studies have suggested that exposure to high levels of environmental allergens (e.g. dust mites, pollen, animal dander, cockroaches) in early life is an important determinant of atopic expression and progression to asthma. Two other asthma risk factors interacting with genetic factors and allergen exposure are tobacco smoke and, possibly, viral infections.

Many factors can trigger an asthma episode, and while some children react to only one trigger, others may develop symptoms upon exposure to many things. Allergens are very important triggers for asthma. Children can have allergies to dust mites, animal dander, pollens and molds, and, more rarely, to foods (e.g. peanut and dairy products). There are several nonallergic triggers, among which upper and lower respiratory tract infections are the most common cause in preschool children. Implicated viruses include rhinoviruses, respiratory syncytial virus, parainfluenza and influenza viruses. Exercise-induced asthma results from breathing cold, dry air which dries out and irritates the lung lining. Children with asthma should not avoid exercise – indeed it is good for them – but they may need to use an asthma medicine before exercise to prevent symptoms (see below). Irritants which can cause breathing problems in children with asthma include air pollution, deodorants, paints, and gases from wood-burning stoves and kerosene heaters. Some medicines (e.g. acetylsalicylic acid), and food additives (e.g. metabisulfite and monosodium glutamate) may provoke asthma, and certain types of weather or changes in humidity may worsen asthma in some children.

Mites and Molds, Pollen and Pets

House dust mites are ubiquitous in home environments and are the major allergic trigger of symptoms in children with asthma. Up to 85% of children with asthma have positive skin tests to dust mites, compared to only 5–30% of nonasthmatic subjects. *Dermatophagoides pteronyssinus* and *D. farinae* are the most common dust mites, the major allergen (Der p 1) being found in their fecal pellets. It has been suggested that the increase in incidence and severity of asthma may be related to the construction of more energy efficient homes which maintain temperature and humidity levels more conducive to the dust mite.

Dust mites are so widespread, however, that their total eradication from the home is impossible. But reducing dust mite exposure helps relieve asthmatic symptoms and reduces medication requirements. Table 1 lists some of the steps that parents can undertake to reduce dust mites in the room requiring the most attention – the child's bedroom.

There has been interest recently in commercial "antidustmite" chemicals (acaricides), such as tannic acid and benzyl benzoate, which are claimed to appreciably lower dust mite levels in the home when applied to

Table 1
Steps to reduce dust mite levels in the bedroom

- Enclose mattress, box spring and pillow in zippered, dust-proof covers
- Remove carpets, fluffy toys and upholstered furniture from the bedroom; the floor surface should be vinyl/hardwood
- Use synthetic pillows and duvets
- Launder bed clothes every 2 weeks at 60°C
- Seal the hot air vent with tape and either use an electric baseboard heater or hot-water heater
- Keep the bedroom door closed as much as possible
- Air condition rooms in the summer to maintain humidity at less than 50%
- ? Acaricides, e.g. tannic acid

[1] A thing of beauty is a joy for ever:
Its loveliness increases; it will never
Pass into nothingness; but still will keep
A bower quiet for us, and a sleep
Full of sweet dreams, and health,
and quiet breathing.

From John Keats, *Endymion*

carpets, furniture and bedding. Marginal improvements in clinical symptoms have been reported, but larger extended studies are required to determine the safety and efficacy of these chemicals and their optimal method of application.

House dust may also contain allergens arising from cockroaches, a common cause of allergic symptoms, particularly in economically deprived urban areas. Eliminating cockroaches in such situations is clearly of particular importance in homes with asthmatic children.

Mold spores, and grass, tree and ragweed pollen are common seasonal triggers of asthma. Because mold spores are smaller than pollen grains they can penetrate the lower airways, and the common mold, *Alternaria*, has indeed been identified as a risk factor for the development of life-threatening asthma. As with pollen, mold spores exist primarily out of doors, but they can also grow indoors in areas of high humidity, such as bathrooms and damp basements. Like house dust mites, pollen and molds cannot be completely avoided. Exposure can be reduced by keeping windows closed during seasons of high mold/pollen production, and by using air conditioning. Humidifiers should be avoided because they harbor and aerosolize mold spores and encourage the proliferation of dust mites. Baths and showers should be bleached once every few months.

Allergens from pets are an important cause of allergic symptoms. Cats are the most frequent source of trouble with potent allergens (e.g. Fel d 1) in dander and saliva. Ideally, pets should be avoided, but if they are kept, they should remain outdoors and washed regularly.

Smoking

Exposure to indoor pollutants and chemical irritants can magnify an allergic patient's reaction to allergens. Children with asthma exposed to parental smoking experience more severe symptoms, have lower lung function, and require more medication. It has also been shown that maternal smoking during pregnancy results in elevated umbilical cord levels of IgE, increased airway reactivity at birth and is a risk factor for the development of atopy and wheezing in early life [2]. Parents of asthmatic children should, therefore, do their utmost to eliminate their child's exposure to secondhand smoke both inside and outside the home. We obviously also require improved counselling of teenagers with asthma, since 20–25% of them are cigarette smokers. Other smoke sources, like wood-burning stoves, and kerosene heaters, which produce sulfur dioxide, should also be avoided.

Treating Asthma

Parents of children with asthma often have an inadequate understanding of the condition, and this has been shown to contribute to the need for hospitalization and in poor compliance. Patient education programs, with emphasis on self-management and promoting a parent-physician partnership, can reduce morbidity and health care costs and enhance the quality of life for children with asthma. The educational issues that need to be addressed are summarized in table 2. Patient education involves not only providing information but should also be aimed at teaching skills (e.g. use of peak flow meters and inhalers; see boxes), changing behavior and establishing parents' confidence that they can manage the asthma. Parents (and children) need to be actively encouraged to participate in the programs, and repetition, regular reassessment of skills and positive reinforcement are vital to success. (The author has prepared an educational booklet on asthma for parents, which is available on request.)

As children mature, they should be encouraged to assume increasing responsibility for their own care. There is also widespread agreement that the education of health care professionals must be strengthened, and national asthma programs have been initiated in several countries to educate parents, school personnel and physicians.

In addition to educational measures, regular follow-up of asthmatic children by a committed physician is essential for optimal care. In this way, the child's progress, response to therapy, compliance and inhalation technique can be reviewed, and lung function monitored. Unfortunately, a significant (and probably increasing) number of asthmatic children use the emergency room as their primary source of care, a practice that precludes ongoing supervision and proper prophylactic measures. Recent studies indicate that up to 70% of patients with asthma do not comply with treatment, a situation which is obviously very far from ideal.

Medication

Asthma medicines can be divided into two groups: preventive (prophylactic) and symptom relieving. Many children will require one of each type of medicine.

Preventive Drugs: Anti-Inflammatories

These decrease the reactivity of the airways and prevent asthma by blocking inflammation. They are used daily and may take anything from 1 to up to 6 weeks to start working effectively. Since they have no bronchodilator activity (see below) they provide no relief of acute asthma symptoms. This group of medications includes both nonsteroids and steroids.

We still don't know precisely how the nonsteroid, *cromolyn sodium*, works, but it is able to inhibit early and late asthmatic reactions when given prior to allergen challenge. In the long-term, it probably provides less effective control of airway reactivity than inhaled steroids. It provides good maintenance therapy for children with mild to moderate asthma. For children with seasonal (e.g. pollen-induced) asthma, administration should begin several weeks prior to the anticipated pollen exposure, and exercise-induced asthma can be prevented by taking it 15–30 min before the exercise, although β_2-agonists are more effective. The major advantage of cromolyn is its virtual freedom from side effects, but it is relatively expensive and the nebulizing solution is rather inconvenient to use.

Although chemically unrelated to cromolyn, *necrodomil* has a very similar clinical profile. Data for children are still limited, and pending further trials, it is difficult to decide on the appropriate niche for this drug in pediatric asthma.

Table 2
Content of parent/patient education program

- Nature, aggravating factors, and prognosis of asthma
- Environmental control measures
- Medication – actions, administration, side effects
- Proper use of inhalation devices
- Acute exacerbations – recognition, treatment, criteria for seeking medical help
- Home monitoring – symptom diaries, peak flow meters

Leukotrienes, which are released from many inflammatory cells, are important mediators in asthma, causing bronchoconstriction, mucus secretion, inflammation and increased vascular permeability. Drugs have recently been developed that can block the actions of leukotrienes, e.g. zileuton, montelukast and zafirlukast. Data regarding their efficacy in childhood asthma are limited, although a recent placebo-controlled study with montelukast in asthmatic children aged 6–14 years did yield encouraging results [3].

Inhaled corticosteroids, because of their high topical anti-inflammatory activity but limited systemic absorption, have revolutionized the management of asthma. They control symptoms effectively and improve lung function in the majority of asthmatic children. Their regular use reduces airway inflammation, bronchial reactivity, and the severity of exercise-induced asthma. Several preparations are now available, including beclomethasone, fluticasone, budesonide, triamcinolone, and flunisolone. Although considerably safer than oral steroids, they are not completely innocuous, particularly when used in higher dosages. Possible side effects include adrenal suppression, growth retardation, osteopenia, decreased bone formation and psychological effects, in addition to topical effects – oropharyngeal candidiasis and dysphonia. Various strategies, such as twice daily administration, use of a spacer device and mouth rinsing after inhalation can reduce these effects.

Symptom-Relieving Medication: Bronchodilators

These drugs relieve asthma symptoms rapidly and prevent exercise from triggering asthma episodes. The most potent bronchodilators available are the β_2-agonists of which there are two main types: short-acting (e.g. albuterol, terbutaline, fenoterol) and long-acting (e.g. salmeterol, formoterol). Short-acting β_2-agonists are used on demand for relief of acute symptoms, i.e. they are "reliever" medicines. Long-acting β_2-agonists should be used regularly (twice daily) in conjunction with higher doses of an inhaled corticosteroid in patients whose symptoms are not controlled by corticosteroids alone, e.g. those who experience symptoms at night or have major problems with exercise-induced asthma.

Although short-acting β_2-agonists are first-line treatment for acute asthma and the prophylaxis of exercise-induced asthma, questions remain concerning their possible use in maintenance treatment.

It remains unclear whether or not β_2-agonists modify the underlying inflammatory process in asthma. Although studies using large doses or long-acting formulations have demonstrated inhibition of the late-phase response and increase in airway responsiveness, these effects are more likely due to functional antagonism of smooth muscle contraction than to anti-inflammatory effects per se.

There are some rather conflicting results on the effects of long-term treatment with β_2-agonists on airway hyperreactivity: studies have reported a decrease, no effect, and increases [4]. That regular use of β_2-agonists might increase airway reactivity is worrying because this might render asthma patients *more* susceptible to asthma exacerbations. Similarly, results investigating regular versus intermittent use of short-acting β_2-agonists have been contradictory, some trials showing that their reg-

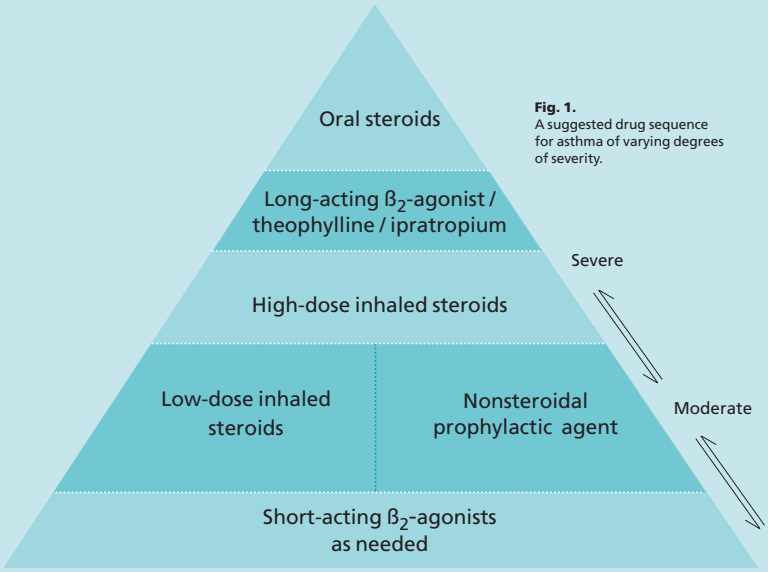


Fig. 1. A suggested drug sequence for asthma of varying degrees of severity.

ular use had a deleterious effect on asthma control, others the reverse. Obviously, further studies are needed to resolve this issue.

Several case-control studies have demonstrated that excessive use of short-acting β_2-agonists is a risk factor for either dying from asthma or having a life-threatening asthma attack. However, evidence from epidemiological studies seems to suggest that most asthma deaths are preventable, and due to overreliance on bronchodilators, resulting in delayed referral to hospital and treatment with systemic steroids, rather than to direct toxic effects of the β_2-agonists themselves.

Clearly the controversies surrounding the long-term use of β_2-agonists have wide-ranging implications. Pending further studies, it seems that (1) short-acting β_2-agonists should be used mainly on demand for the relief of acute symptoms and (2) dependency on short-acting β_2-agonists should be considered an indication to introduce/increase anti-inflammatory therapy.

Fortunately, oral steroids are rarely necessary today in the long-term treatment of pediatric asthma. If they are needed, small doses of prednisone/prednisolone are given, on alternate days if possible. However, short courses (5–7 days) are extremely effective in treating acute exacerbations of asthma and may reduce the need for hospitalization. Their use may cause transient adrenal suppression, but recovery is rapid after their withdrawal.

Guidelines for Drug Use in Asthma

Given the public health and economic impact of asthma, medical organizations in several countries have formulated guidelines for its diagnosis and treatment [1, 5]. Emphasis has been placed on the chronic nature of asthma and on the early introduction of prophylactic medication to reduce airway inflammation and reactivity. Thus, although the intermittent use of a β_2-agonist may suffice in children with mild and infrequent asthmatic symptoms, prophylaxis with an anti-inflammatory medication is recommended when the symptoms are more frequent and severe (fig. 1). Since the prophylactic agents have no bronchodilator acitivity, an asthmatic child must at all times have access to a short-acting β_2-agonist (and oral steroids for some patients) to treat acute symptoms.

Into the Emergency Room

As already indicated, if we can improve patient and parent awareness of the nature of asthma and how to control and treat it adequately on a day-to-day basis, we should be able to prevent, or at least ameliorate, the acute exacerbations of asthma that bring the child into the hospital. Acute asthma is a potentially life-threatening medical emergency, although deaths are, fortunately, relatively rare.

Regardless of the precipitating factor, the central event is widespread airway obstruction that results from bronchial smooth muscle spasm, inflammation and mucus plugging. Airway obstruction leads to increased airway resistance, reduced flow rates, gas trapping and pulmonary overdistension. Severe asthma exacerbations may lead to respiratory failure and can be potentially life-threatening.

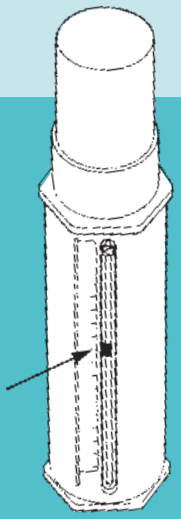
Children with severe acute asthma have to be treated expeditiously and observed closely. The goals of treatment are relief of the hypoxemia, quick reversal of airway obstruction, and prevention of early relapse. Usually, oxygen is supplied by face mask together with nebulized medications, and intravenous fluids may be necessary to supplement fluid intake and provide access for the administration of drugs. Short-acting β_2-agonists are the most effective initial treatment, usually supplied by a nebulizer. Anticholinergic agents, theophylline, or corticosteroids may be used additionally, depending on the individual situation – since children with asthma at all levels of severity may be admitted to the emergency room, the intensity and duration of therapy will vary accordingly – the therapeutic approach has to be flexible.

After appropriate treatment in the emergency room, up to 75% of children with acute asthma can be discharged home, i.e., they do not require hospitalization. Before the child is discharged, it should be established that his/her condition is stable, the child's inhaler technique should be checked and the importance of compliance with medications stressed. At home, inhaled bronchodilators should be used regularly over the next few days in addition to, for most children, a short course of oral steroids. The child's general practictioner should then review and reassess the situation soon after the emergency event, providing further education, ensuring proper long-term prophylaxis and ongoing supervision. This way, future acute exacerbations of the disease can, hopefully, be prevented.

Box 1

The peak flow meter

Peak flow meters can be used regularly at home to find out how well a child's asthma is controlled and how well medicines are working.
Standing up, the child takes a deep breath, places the mouthpiece in the mouth, closes the mouth around it and then blows out fast and hard. This is repeated twice, and the best of the three readings is noted.
By recording the peak flow rate on a diary card each morning and evening, the child's "personal best value" can be ascertained, i.e. the highest peak flow number when the asthma is well-controlled. When the asthma is getting out of control, the peak flow rates may begin to drop hours or even days before symptoms develop: rates 50–80% of the personal best are warning signs, values below 50% signal a medical alert. By taking extra medication before symptoms appear, an asthma attack may be prevented.

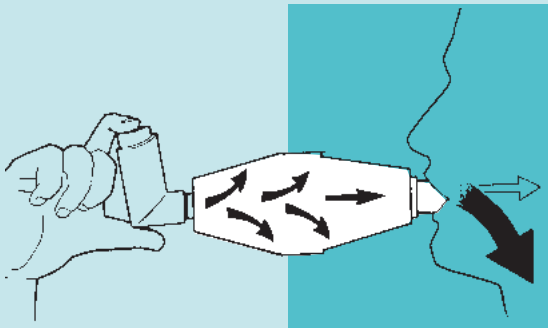


Mini-Wright peak flow meter

Box 2

Inhalers

Metered-dose inhalers deliver the medicine as a spray and work best when used with a spacer device which ensures that more medicine reaches the lung and less impacts on the throat. Metered-dose inhalers are sometimes just called "inhalers" or "puffers."
Dry-powder inhalers are convenient because they are small and do not require a spacer device. To use them, air needs to be sucked in rapidly.
Nebulizers deliver the medicine as a fine mist. They are often used in the clinic or emergency room or at home when children are too young to use a metered-dose inhaler. A treatment takes about 15 min.

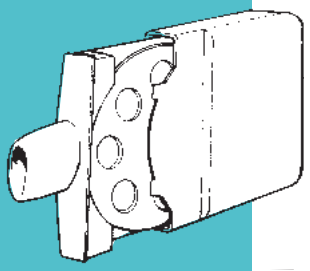


Metered-dose inhaler with spacer

How a metered-dose inhaler is used

1 The inhaler is shaken and attached to the spacer.
2 The child breathes out fully and closes his/her lips around the spacer mouthpiece.
3 The spacer is loaded with one puff of aerosol.
4 The child inspires slowly and deeply.
5 The breath is held for 10 seconds (or as long as possible).
6 If more than one puff or more than one medication is required, the child must wait 1 minute before inhaling again.

Some examples of dry-powder inhalers



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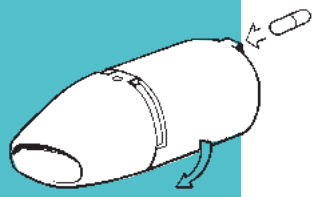

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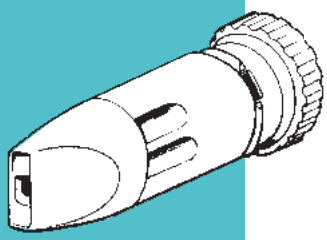

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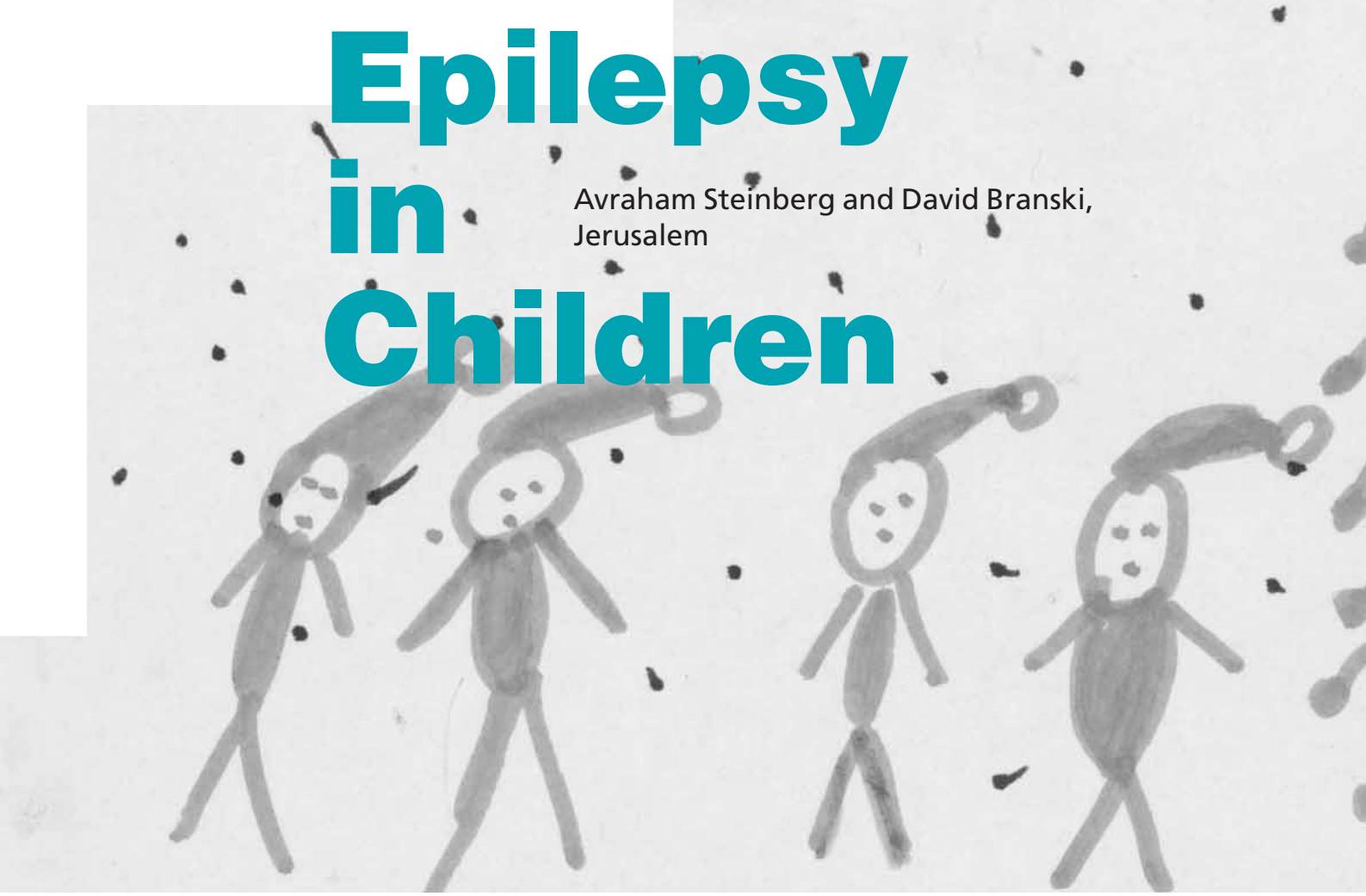
Epilepsy in Children

Avraham Steinberg and David Branski, Jerusalem



A Brief History of Epilepsy

Epilepsy was known to most ancient societies and described by both their physicians and lay people. The term 'epilepsy' is probably derived from the Greek word *epilambanein*, meaning sudden seizure, or from *epi*, meaning above, and *lesis*, meaning affliction, i.e. it is an affliction of the head. Due to the frightening experience of loss of consciousness, cessation of breathing, and violent movements, the ancients referred to this disorder as the disease of demons or "the holy disease" – a title Hippocrates gave to one of his works, and a term subsequently used by many physicians. They assumed that the affliction had supernatural causes, or came from the head, the "holy site." The stigma attached to the disease led to it being referred to as the great illness (*grand mal*) or the small illness (*petit mal*) without further specifications (fig. 1).

Epilepsy has been portrayed in many different ways down the ages. Epileptics were thought to have unique and negative personalities who would eventually become mentally retarded and behave abnormally. For these and other reasons they were ostracized from society, restricted in their activities, considered to be fools and mad. The illness was judged to be shameful, a form of curse.

A number of famous historical figures suffered from epilepsy: Socrates, Caesar, Dostoevski, Lord Byron, Berlioz, and others. Interpreters assume that the biblical characters Balaam and King Saul were afflicted by the disease.

Theories for the origins and causes of epilepsy have been legion. Greek physicians believed that it was caused by one of the four body humors, and also that gases produced from ingested foods ascend through the veins to the brain causing seizures there. Epilepsy has been attributed to violence, demons or magic, to lunar or other celestial influences, or to various alchemical actions. Some people thought that sexual intercourse caused epileptic seizures leading them to suggest castration as a cure. Others have related seizures – or their prevention – to menstruation and/or pregnancy. Epileptic fits have also been ascribed to various sensory changes in vision or hearing, and to severe pain.

It was not until the 19th century that seizure disorders were accorded the status of a medical illness. In 1860, The National Hospital for the Paralyzed and Epileptic was opened in London to care only for apoplectic and epileptic patients. The first medical textbook on epilepsy was published in 1881, while rapid advances in understanding epilepsy came with the development by Hans Berger in 1929 of a machine to measure the electrical activity of the brain – the electroencephelograph (EEG).

The EEG

The EEG consists of a set of electrodes which are placed on designated spots on the scalp and are connected by wires to a machine which greatly magnifies the electric brain waves and traces them out on paper. The EEG recording is the single most valuable laboratory test in the majority of children with seizures and is required in any case where epilepsy is suspected, being performed as close to the event as possible.

The EEG supplies information about several aspects of a child's brain function. It aids in diagnosis and classification, helping to define the particular epilepsy type and thus the treatment decision. It also provides information about the background activity of the brain and possible underlying etiological factors.

Long-term video-EEG monitoring is helpful for intractable epilepsies, or in cases where there is poor correlation between the history of seizures and the clinical assessment and the routine EEG trace.

Other imaging modalities, such as computed tomography or magnetic resonance imaging are utilized in specific situations, such as focal seizures, where the disease is intractable, and where the history and/or physical examination suggest the possibility of structural brain abnormalities (e.g. tumor, malformation, degeneration).

The Current Definition of Epilepsy

Epilepsy is a chronic disorder of the brain, manifested by parox-

Fig. 1. This 19th-century engraving shows a man suffering a grand mal ("great evil/sickness") seizure.

ysmal electrical discharges, which cause recurrent seizures of various clinical presentations. Such individual episodes are called seizures, fits, or convulsions.

Epilepsy is not a unitary disease, but a group of disorders which differ significantly in etiology, the form of the electrical discharges in the brain, the clinical manifestation of the seizures, hereditary predisposition, response to treatment, prognosis, and the various associated manifestations.

There are several classifications of epilepsy based upon the EEG abnormality, age of onset, or the clinical manifestation. Table 1 summarizes the current classifications. These are tentative, and will evolve and become modified as we learn more about the disease.

How Frequent and Why

It has been estimated that the chance of having at least one seizure by the age of about 80 is 8%. Fifty percent of individuals who have seizures have the first (and sometimes the only) one during childhood or adolescence.

The annual incidence of newly diagnosed epilepsy in children under 20 years of age is 0.5–0.8/1,000 per year. Between 3.4 and 5.7 out of 1,000 children are taking medication for active epilepsy. The overall risk of developing epilepsy during childhood or adolescence is about 1%.

Different epilepsies have different etiologies. In general there are three major underlying causes: genetic predisposition, neuropathological changes, and chemicophysiological alterations in the nerve cell and its connections. However, despite the many advances made in this field, in only a small number of cases can the cause of epilepsy be identified – most diagnoses are for idiopathic or primary epilepsy (fig. 2).

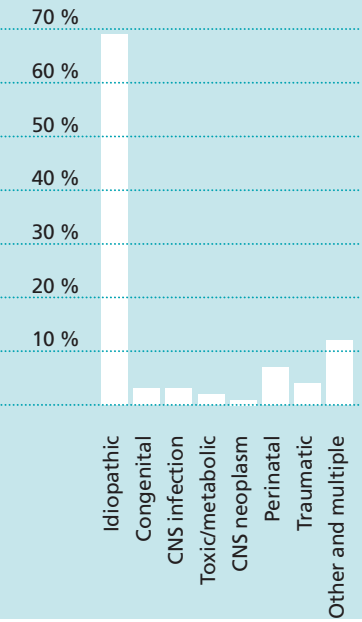


Fig. 2.
Attributed causes for epilepsy

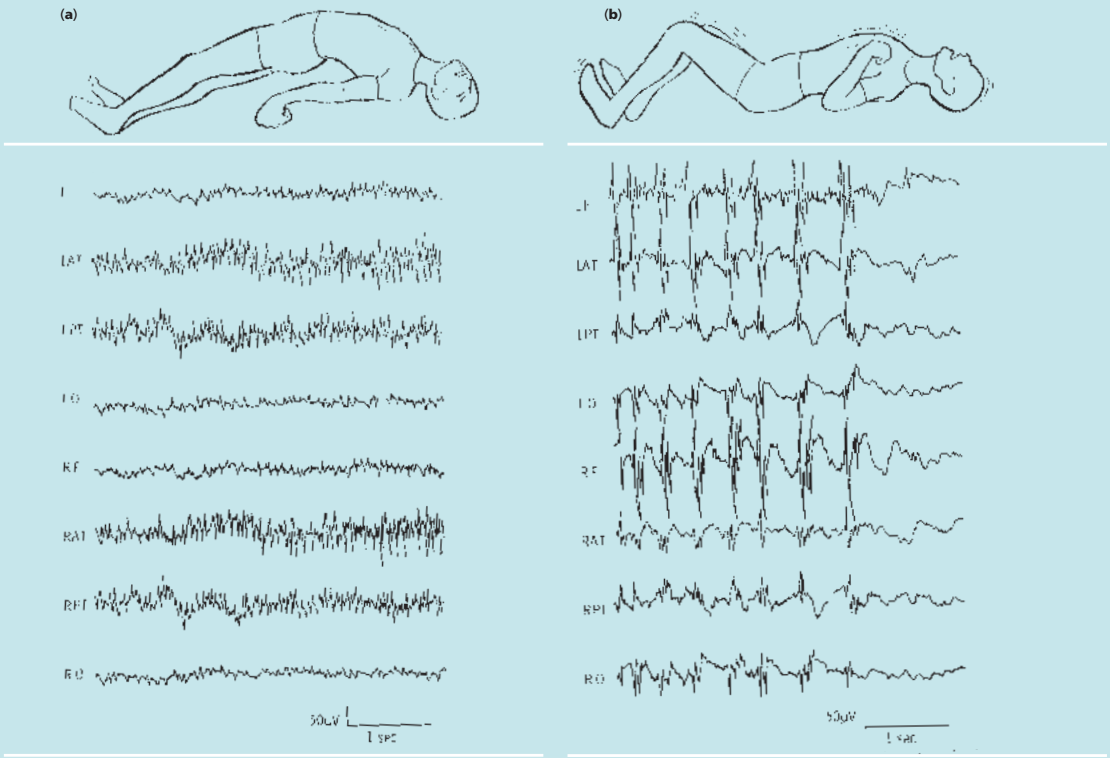


Fig. 3. Patient during the tonic (**a**) and clonic (**b**) phases of a grand mal seizure with the accompanying characteristic EEG traces.

What Happens in an Epileptic Seizure?

Clinical seizures or fits are caused by acute, rapid, and sudden discharges of electrical activity from the cerebral cortex or from deep cerebral nuclei in the brain. This abnormal activity may be generated from one or more sites in the brain, or from the whole brain. The symptoms are very variable and may include loss or change of consciousness, frothing in the mouth, abnormal behavior, abnormal sensations, rapid focal or generalized body movements, focal or generalized muscle spasms, falling to the ground, vomiting, loss of urine and/or bowel control, salivation from the mouth. A seizure may last anything from several seconds to hours. The abnormalities are chronic, recurrent, sudden, and usually unpredictable. Between episodes a child may act perfectly normally or demonstrate various mental abnormalities.

Seizure episodes can be divided into three stages: (1) the aura, during which the imminent seizure is sensed, (2) the clinical seizure (ictus), and (3) the postictal period, which may involve tiredness, confusion, and various unusual sensations.

The Forms of Epileptic Events

Grand mal or primary generalized tonic-clonic epilepsy

In this form of epilepsy, the child typically loses consciousness and then has a convulsion consisting of tightening of all the muscles (tonic phase) followed by jerking of the arms and legs (clonic phase) (fig. 3). The seizure may be accompanied by rolling up of the eyes, turning pale or blue, wetting or soiling, and biting the tongue or cheek. Afterwards the child may be sleepy, confused, exhausted, and complain of a headache.

During the attack, the EEG shows abnormal discharges of spikes and sharp waves. It may also be abnormal between clinical attacks.

Petit mal or typical absence epilepsy

This is an epileptic form usually seen only in children, disappearing in adult life. The seizures consist of a sudden brief loss of contact with the environment (absence) lasting from a few seconds to a minute, without falling to the ground. The child appears to be day-dreaming or staring, may blink, and there may be slight jerking of the arms and legs. After the interruption in consciousness, the child will continue doing whatever he or she was doing just before the attack. Such seizures may occur many times during the day.

The EEG (fig. 4) is pathognomic, i.e. it has a unique pattern (in this case, three general complexes per second, spike and slow wave). The clinical and electrical attacks can be induced by overbreathing.

Partial complex, psychomotor, or temporal lobe epilepsy

There are various ways in which this type of epilepsy manifests itself. The child may perform repeated complex movements (automatic behavior) associated with a clouding of consciousness. Or there may be atypical absences, longer than those associated with petit mal, and usually followed by a postictal state of confusion. Or the child will experience tonic-

Table 1. Classifications of epilepsy

1. **Etiological classification**
   a. Primary or idiopathic
      These have no known etiology
   b. Secondary or symptomatic
      A cause for the recurrent episodes can be idenified

2. **International League against Epilepsy classification**
   a. Partial epilepsies
      Characterized by a focal electric discharge in the brain and/or focal clinical manifestation of the fits
      A seizure may begin focally and evolve into a general one (secondary generalization)
      These seizures can be further subdivided:
      i. Simple – no change in consciousness
      ii. Complex – change in or loss of consciousness
   b. Primary generalized epilepsies
      General electric discharges in the brain and generalized clinical fits
   c. Unilateral seizures
   d. Unclassified seizures

   The separation into partial and generalized seizures is somewhat artificial because a continuum between the two entities has been recognized.

3. **A syndromatic approach**
   This is perhaps the most appropriate means of classification in pediatric epilepsy. The description and classification covers a host of characteristics such as clinical manifestation, electrical pattern, etiology, and response to treatment.

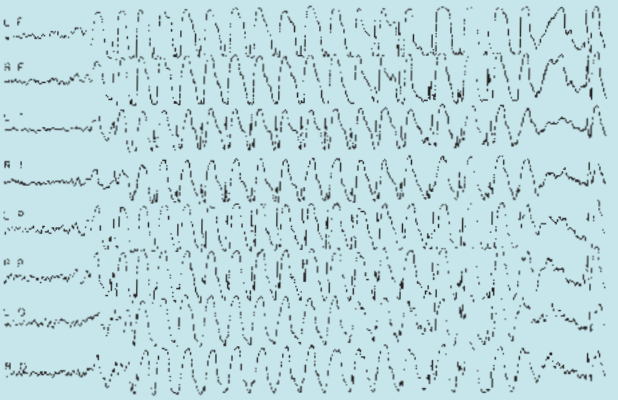


Fig. 4. Absence seizures. At far left are normal brain wave patterns from eight sites in the brain. They are followed by the steep "spike and wave" EEG pattern associated with absence seizures.

clonic movements with a focal element, occasionally evolving into secondary generalization.

Seizure episodes are occasionally preceded by a warning sign, the aura. An aura may comprise strange feelings in the abdomen, spots before the eyes, the sensation of unusual tastes or odors, or anxiety.

The origin of this type of epilepsy is usually an abnormality in the temporal lobe(s) of the brain.

West syndrome, or infantile spasms

This type of epilepsy is restricted to infants. It is characterized by a triad of features: brief spasms resembling a greeting position (salaam attacks), which last for several seconds and occur in clusters, especially on waking; arrested psychomotor development leading to mental retardation, and a pathognomic EEG pattern called 'hypsarrhythmia' (fig. 5).

Myoclonic epilepsies of childhood, or Lennox-Gastaut syndrome

This is an epilepsy of early childhood. A variety of seizure types may occur – tonic-clonic, myoclonic, atypical absences, and/or drop attacks – combined with a typical abnormality in the EEG tracing, severe mental retardation and severe behavioral problems.

Unfortunately, we rarely know what triggers a seizure and there are usually no warning signs. But some factors can exacerbate the chances of an event, such as the abrupt withdrawal of an anticonvulsive drug, a high temperature, head trauma, fatigue, or lack of sleep.

Diagnosis and Treatment

The most important elements in the diagnosis are the history (see table 2) and the EEG tracing. The ultimate goal of epilepsy management is to assist the patient to lead a full, high-quality and ultimately productive life.

Explanation, reassurance and psychological support are very important considerations in the management of epileptic patients. In the case of very young children who suffer from epilepsy, it is the parents who need such support. They frequently imagine the worst scenario concerning their child's future: severe retardation, difficulties at school, problems acquiring a profession. The parents also fear the emotional and economic burden they might face. In addition there are the social stigmas related to epilepsy, and occasionally there is the feeling of guilt on the part of the parents. In many epilepsy clinics and units, support services such as social workers, psychologists and educators are on hand to help parents better understand the nature of the illness, the varied outcomes of the different types of epilepsy, modern understanding of the illness and the improved therapeutic options available.

Older children who suffer from epilepsy have their own anxieties, fears and imagination of what is happening to them and what will happen in the future. Many parents prefer to conceal the information from children to spare them the anxiety connected with the diagnosis. However, intelligent children will sense the deception and this only augments their fears. Thus both parents and children require counselling, reassurance, and psychosocial support.

One important measure in the treatment of epilepsy is to prevent and correct precipitating and exacerbating factors. However, the cornerstone of epilepsy management is the prescription of anticonvulsive drugs. Where these fail, as they will occasionally, we have to resort to special treatments such as dietary changes or, rarely, neurosurgery.

Ideally, anticonvulsives will prevent seizures and stop episodes when they do occur. We usually start with one drug, chosen according to the clinical and EEG characteristics. It is important to assure compliance, with timing and administration depending on the specific pharmacological characteristics of the drug. We often encounter compliance problems with children. Some children find it difficult to swallow tablets, and thus need the medication in the form of a syrup or chewable pills. However, not all anticonvulsants are available in such forms. Older children can sometimes be rebellious and refuse to take the medications in a regular and scheduled manner. Unfortunately, this may worsen their seizure disorder.

The initial drug is used until there is a therapeutic effect or signs of toxicity appear. Obviously, where the drug is ineffective or demonstrates toxicity or side-effects it has to be changed. It is crucial, however, that anticonvulsive medication is not stopped abruptly – doing so may provoke or exacerbate seizures.

Fortunately, a wide range of anticonvulsive medications are now on the market, the newer formulations having a broader range of indications, lower toxicity, and eliciting fewer side effects.

Once complete seizure control has been achieved the drug should be taken for 2–3 years and can then be gradually discontinued.

There may be several reasons why anticonvulsive medication is ineffective: noncompliance, incorrect classification of the seizure type, wrong choice of drug, over- or under-dosage or drug interactions, failure to recognize other diseases or triggering and exacerbating conditions, failure to deal with emotional disturbances.

If all these considerations are ruled out and the child suffers from seizures that disrupt his or her life for more than a year, then the diagnosis has to be one of intractable epilepsy and alternative treatments considered. These may include immunoglobulins, steroids, a ketogenic diet, vagal stimulation, or neurosurgery.

Children with continued seizures of focal onset are potential candidates for surgery. There are three types of epilepsy surgery: (1) removal of a defined lesion in the brain – e.g. tumor, cyst, vascular malformation – responsible for the repeated seizures; (2) epileptogenic surgery, the removal of the brain area generating the abnormal electric activity giving rise to the seizures; (3) corpus callostomy, surgical removal of most of the corpus callosum to minimize the spread of abnormal electrical activity from one hemisphere to the other.

The Long-Term Outlook

Most epileptic patients live a close to normal life, without any mental or social problems. However, even the "easy" cases with good outcomes require education

Table 2. Taking a history for epilepsy

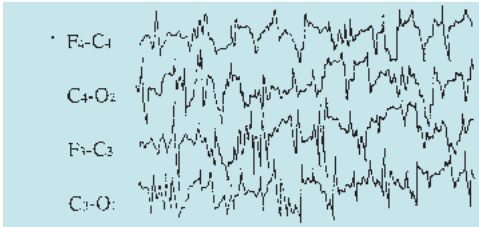
Many of the answers to the following questions will have to be provided by eyewitnesses to the event(s), especially in the case of young children.

1. Were there provocative factors, such as fever, head trauma, or anticonvulsant drug withdrawal?
2. Is there any underlying medical or neurological illness?
3. Were there warning (aura) experiences?
4. How did the seizure manifest itself?
   Form?
   Duration?
   Associated symptoms?
5. What is the frequency and duration of attacks?
6. Were there any associated behavioral abnormalities?
7. What was the age at onset of the seizures?
8. Is the patient taking anticonvulsive medication?
9. Are the seizures well-controlled by medication?
10. Is there a relevant family history concerning epilepsy?

and psychological support during the period of treatment. Both parents and children need to be reassured that the treatment is both needed and beneficial. Overall, approximately 70–80% of children who develop epilepsy will enter remission, and will subsequently stop having seizures.

A small number of epileptics do suffer from mental deficiency or have social conduct problems. Some are intractable – we cannot adequately control their seizures with anticonvulsant medication.

Epileptic children may experience learning disabilities, attention disorder and behavioral problems, either related to the underlying brain pathology, to inadequate seizure control, or to the effect of medications. They may also suffer from social stigma and discrimination, but if the disorder is diagnosed early and treated adequately, we will be able to ensure that the great majority of epileptic children will live a normal childhood with all its concomitant joys, fears and lessons.



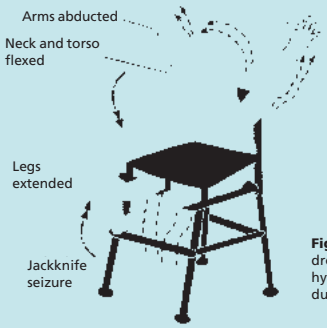


Fig. 5. Infantile spasms (West syndrome). The EEG shows the hypsarrhythmia typical of the pattern during such an attack.

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What to Do for a Person with a Grand Mal Attack

Act as calmly as possible

Be assured that despite the frightening experience witnessing a seizure, this is not a life-threatening situation if the duration of the fit is short (i.e. a few minutes).
An exact description of the clinical details of the seizure is of great importance for diagnosis.

Protect the patient from injury

Help the patient lie down on the floor or a bed, away from edges and sharp corners, turn the face to one side, so that vomit will not be aspirated.
There is no need to try to force open the mouth of someone during the tonic phase of the attack.
There is no need for mouth-to-mouth resuscitation.
There is no use e.g. pouring water over, smacking or pinching the patient.

In the case of a first seizure

Get the patient to hospital as quickly as possible.

In the case of known epilepsy

If the acute attack ceases within seconds or even within a few minutes, there is no need to rush to hospital – just notify the patient's physician.
A patient experienced with such events may use a rectal form of valium (i.e. a Stesolid rectal tube), which will either abort or at least shorten the attack.

After the acute seizure

Patients usually fall asleep – let them sleep as long as they need!

Childhood Cancer

Each year in Switzerland, about 180 children, aged 0–16 years, are newly diagnosed with cancer. Statistically, this means that pediatric cancers are rare, but for the child with the disease, its parents, and other family members, the diagnosis announces a significant deviation from a 'normal' childhood punctuated by the usual scrapes and knocks, mild infectious diseases, and the odd broken bone. Months, sometimes years of intensive monitoring and treatment lie ahead, pain and anxiety met and, hopefully, overcome. While many – the majority in fact – of the children will be cured and go on to lead healthy adult lives, for a few, the disease is fatal. They, and those close to them, will be forced to confront the issues of mortality and loss that most of us tend to push to one side until well into adulthood. To learn more about the current treatment of children with cancer, the Gazette's editors visited ***Dr. H. Plüss***, senior consultant in pediatric oncology at the Zürich Children's Hospital. The following article is based on our discussion.

There are quite large regional differences in the incidence of the various childhood cancers worldwide. Switzerland is fairly typical of the industrialized countries of the northern hemisphere: about one-third of pediatric cancers are leukemias and one-quarter brain tumors. Turkey has a higher incidence of Hodgkin's disease than other European countries (no one yet knows why), in Pakistan non-Hodgkin's lymphoma is much more prevalent, retinoblastoma is very common in Brazil.

This spectrum might well be changing though as AIDS makes its impact. Dr. Plüss said that in Zürich, an increase in non-Hodgkin's lymphoma among children, almost certainly attributable to HIV infection, was already noticeable, and in other countries, like the United States, different cancers are now being seen in HIV+ children.

Cancer is fundamentally a genetic disease – any chromosome damage might lead cells to lose their specialized, differentiated state and break away from the normal constraints on growth. Obviously, 'something' has to cause the mutations, or prevent their normal repair, but there is still intense debate about which, and to what extent, environmental factors (like radiation, chemicals, etc.) are carcinogenic. Our immune systems are continuously monitoring our entire bodies for, and disposing of, cancer cells. Dr. Plüss made the interesting observation that while about one-sixth of Switzerland's residents are foreigners, notably more than one-sixth of his patients were born outside Switzerland, suggesting perhaps that they lack immunity to certain factors, like viruses, that may be in part responsible for some cancers.

Treating Cancer Today

Cancer is scary – it conjures up visions of weight loss and pain, of treatments with very unpleasant side effects and, of course, death. But the last couple of decades have witnessed impressive improvements in the treatment of some childhood cancers – both in terms of cure rate and the quality of life during and after treatment. For many children, the diagnosis of cancer is no longer a death sentence.

Today, 80% of children with leukemia will be cured. Similar rates are obtained with Wilm's tumor (a kidney cancer) and Hodgkin's disease. Spectacular success has been made in the treatment of Burkitt's lymphoma, a cancer especially prevalent in Africa, and associated with Epstein-Barr virus infection. In the 1980s, it was fatal. Now, with a short 4-month treatment, which includes, among other drugs, a very high 24-hour dose of methotrexate (immediately followed by an antidote to counteract the side effects of the drug), the cure rate is 80% and relapse is rare. Indeed, if there is no relapse within 2 years, the patient is considered totally cured. At the other end of the spectrum though, neuroblastoma, if it occurs after 1 year of age, remains almost incurable. Brain tumors tend to fall somewhere between these extremes: about 50% of children with such tumors will survive, but very often with severe brain damage, in part due to the tumor itself, in part to the side effects of surgery and radiation therapy.

Retinoblastoma is a classic 'model' cancer, induced in a well-defined two-step process involving the deletion of an antioncogene. It occurs in very young children, usually in the first year of life, and is rare – the Zürich hospital sees perhaps one new patient a year. The cure rate is around 90%, but very often the patient will go on to develop a second, unrelated cancer later in life. This highlights the necessity, in certain cases, for long-term (sometimes lifetime) monitoring of former childhood cancer patients.

The predominant treatment mode for pediatric cancers is chemotherapy, and young children tolerate it extremely well, in part perhaps because they are not as much prey to the psychological anxieties of adults that provoke negative drug reactions.

Unless there is no other choice, radiotherapy is avoided for treating children's cancers. It is too often associated with long-term, not easily reversed, consequences such as brain damage, growth retardation, and secondary cancers. Bone marrow transplantation is also not a common method of treatment, although in Zürich, Basel and Geneva there are beds for pediatric bone marrow transplantation, and autologous trans-

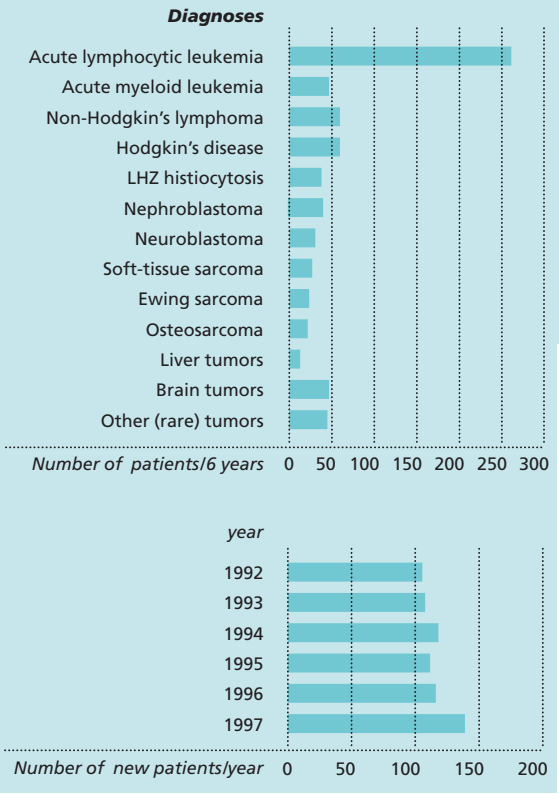

Fig. 1. Children with malignant diseases treated on international or national protocols in Switzerland 1992 – 1997

plantation may be undertaken in, for example, cases of acute myelocytic leukemia and neuroblastoma.

The Swiss Pediatric Oncology Group (SPOG)

Pediatric oncology, like oncology in general, is a relatively recent medical discipline, a post-Second World War phenomenon, and several notable physicians have contributed to the study of cancer in Switzerland. Conrad Gasser working in Zürich was the first to cure a leukemia in this country, in a child, in 1952, and S. Barandun, an immunologist in Bern, who worked on myeloma, was gifted in raising large sums of money to invest in oncology research and to support young oncologists. Dr. Plüss cofounded SPOG in 1976, the main aim of the organization being to coordinate the pediatric oncology groups at the various major Swiss hospitals in developing national protocols for treating childhood cancer. The first such protocol was for acute leukemias, and was used until 1990. They also developed protocols for brain tumors.

The 1990s, however, have witnessed an increasing return to the kind of international collaboration that existed in the immediate post-war years (most Swiss oncologists at that time being trained abroad). Today, trials of new protocols need such large sample sizes that a relatively small country like Switzerland cannot possibly conduct one alone. Oncologists in Zürich not only have close contact with American colleagues, but are closely involved with their German-speaking counterparts in Germany and Austria on leukemia and lymphoma initiatives, in which the Germans, in particular, have a long tradition.

In Switzerland, the 'young' discipline of pediatric oncology has encountered serious financial and administrative constraints within the fairly rigid and slow-to-evolve university structures. The Department of Pediatric Oncology has no secretariat and money is hard to come by. The so-called 'soft' money that backs cancer research and positions in other countries is largely lacking in Switzerland – banks, for example don't, yet, support medical research, considering it to be a state problem. Groups like SPOG, and its umbrella organisation, SIAK (The Swiss Institute for Applied Cancer Research), are therefore crucial in drawing the attention of both politicians and the general public to the needs of clinical cancer research, such as finding out more about the long-term consequences of childhood cancer, and what it may be able to achieve if adequately supported.

Mind As Well As Body

Cancer isn't only a physical disease, it causes great emotional stress too. Not only for the patients, but also for their relatives and friends. Dr. Plüss described how much doctors' attitudes have changed towards these 'significant others' over the years. In Zürich, the doctors inform the parents fully about the diagnosis, treatment procedures, and possible outcomes. Whereas in the past the child being treated would be separated from its siblings, today the whole family is encouraged to come with the child to the hospital on the day she or he has an appointment, and they are allowed into the treatment rooms while an examination is underway, blood samples and biopsies are taken, and so on. This is one way to offset the neglect that brothers and sisters may experience when their parents invest so much time and energy in the sick child.

Psychologists are an important component of the pediatric oncology team, always available for the children themselves and members of their families. And psychologists are also there for the nurses. A pediatric oncology ward is no soft assignment, and while the doctors are in a position to place a certain amount of necessary distance between themselves and their patients, it is the nurses who must be there around the clock supporting patients and relatives in sometimes distressing situations. Although most terminally ill children are cared for at home, it is usually the nurses, not the doctors, who are present when an inpatient dies.

Both parents and children, when they're old enough, are allowed a say in the treatment process. Disagreements between them and the doctors are, however, rare. The most 'difficult' group of patients – from the doctors' perspective – are adolescents. In fact they form only a small percentage of pediatric cancer cases, most childhood malignancies, with the exception of brain tumors, occurring in the first 4 years of life. Adolescents, not unlike their healthy counterparts perhaps, can get into conflicts with both their parents and physicians that may lead, occasionally, to their giving up the recommended treatment altogether. Whatever the doctor's opinion in such a situation, these wishes are respected.

In Switzerland, as elsewhere, associations of parents of children with cancer play an important role on the pediatric oncology scene. They liaise with hospital staff and can often provide doctors with valuable feedback about their care practices. They provide a support network among parents, sharing knowledge, advice and good old-fashioned sympathy. And, in Switzerland at least, they have in recent years become very active in fund-raising, not only for clinical services, but for basic research too.

The Future

When we asked Dr. Plüss to gaze into a crystal ball and speculate how the treatment of childhood cancer might develop in the near future, he came up with two main observations. First is the need to find therapies and cures for the still intractable diseases, like glioblastoma, which, although more common in adults than in children, is virtually always fatal. Second, and not entirely independent of the first, is the requirement for new treatment modalities, especially for those diseases where the success rates are now quite high, but clearly plateauing. Chemotherapy, as Dr. Plüss put it, is now so sophisticated, not much more can be expected of it. What are the possibilities? In the shorter term, immunotherapy seems to offer the most promise. And then there's gene therapy. Zürich is participating in trials for glioblastoma, which involve introducing antiviral agents into cancer cells in the brain. Nevertheless, although newsworthy and technically exciting, gene therapy is, Dr. Plüss believes, still a basic research technique, for which many problems have yet to be solved – its widespread application in clinical settings is still several years off.

So, perhaps we could say that the 'young' discipline of pediatric oncology has reached early middle age. It can look back on several decades of achievement but, not complacent, must now combine the lessons of the past with contemporary innovations in order to secure for today's, and tomorrow's, children with cancer a bright and healthy future.

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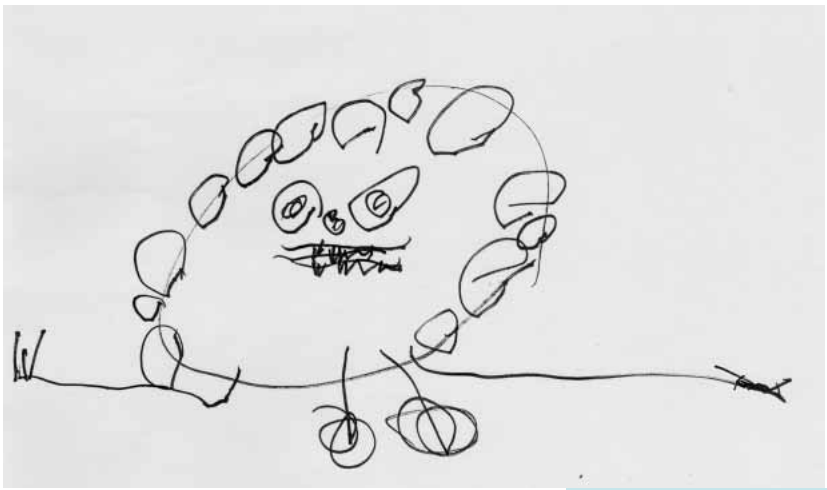

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