

## Book

### Prejudice in a portrayal of Huntington's disease

Along the northeast shores of Lake Maracaibo, in Venezuela, there are communities with a staggeringly high prevalence of Huntington's disease. They live as outcasts, in abject poverty, against a backdrop of the skyscrapers of Maracaibo itself. These people are descended from a woman who lived in Venezuela in the early 19th century; and her genetic inheritance has now passed through ten generations.

In Barranquitas, for example, families affected by Huntington's disease are ostracised from the main town; they live in corrugated iron shacks with earth floors. When you walk through their enclave you soon see the entire spectrum of this terrible disease. It ranges from apparently normal children, to a few affected children, to obviously affected adolescents and young adults, and to bedridden men and women in their 30s and 40s who are awaiting death. Yet these families possess great dignity. Their worn clothes are spotlessly clean; the warmth of the invitation into their homes is genuine; and their offer of refreshments is more than a courtesy. This community scrapes a living by fishing in the lake, but fish stocks have dwindled over the years and survival is precarious. They have few health-care facilities. Yet it was samples from these people's DNA that showed, in 1983, where the genetic abnormality was located (chromosome 4) in Huntington's disease; and it was these same people's DNA that was used to identify, in 1993, the precise nature of the genetic defect (a repeating CAG sequence).

The contrast between the worlds of these families with Huntington's disease around Lake Maracaibo, and that of the hero—or perhaps anti-hero—of Ian McEwan's novel *Saturday*, could hardly be greater. The novel follows 24 hours in the life of a London neurosurgeon named Henry

Perowne. As well as being wealthy and successful, Perowne is a self-serving, self-satisfied, arrogant, name-dropper. During the course of these 24 hours, Perowne has an altercation with a man—the villain of the book—known only as Baxter. We are introduced to Baxter as he runs from a "gentleman's club" in London, accompanied by two thugs. He is short, dressed in a black

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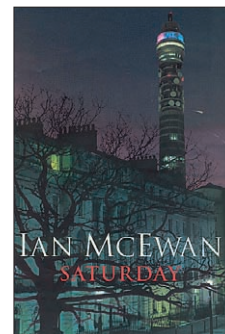
suit, and everything about him is portrayed as sinister: "He's a fidgety, small faced young man with thick eyebrows and dark brown hair, razored close to the skull. The mouth is set bulbously adding to the effect of a muzzle. The general simian air is compounded by sloping shoulders and the built-up trapezoids suggest time in the gym, compensating for his height perhaps. He gives an impression of fretful impatience, destructive energy, waiting to be released."

The critical moment in the book occurs when Perowne's sleek Mercedes sideswipes Baxter's BMW. Baxter's car loses a wing mirror but Perowne's only suffers minor surface scuffing. Perowne's arrogant insouciance—he is eager to get to his regularly scheduled Saturday squash game—ignites Baxter's ire. Baxter wants money—and satisfaction—not insurance particulars. Offended by Perowne's high-handedness, Baxter strikes a blow to his chest. But Perowne hits back even more dangerously: "Your father had it and now you have it too." He has recognised that Baxter has Huntington's disease.

As the book progresses, we learn that Baxter is aware of his symptoms; he knows there is no cure; and he understands his fate. Like his father, he will develop progressively incapacitating chorea and increasingly severe cognitive impairment. The disease will ultimately kill him. Later in the day, Baxter appears at Perowne's home with a knife and threatens his family.

Throughout the book, McEwan attributes Baxter's movements, thoughts, and actions to his Huntington's disease. His volatility, lability, and his emotional freedom to be dangerous and murderous are all features of a man who has nothing to lose. He is dying anyway. His "simian air" is melded into a face constantly beset by movements; the failure of the tracking movements of his eyes are ridiculed—even though Perowne describes these as manifestations of lost neuronal connections. And even when Baxter becomes enraptured with a poem, it is held up as an example of emotional and dangerous instability: "Baxter is hovering behind them, making frenetic little dips of his body. He's becoming manic, he's tripping over his words, and shifting weight rapidly from one foot to the other . . . Now Baxter nips forwards and seizes it [the poetry book], waves it in the air, as if he could shake meaning from it. 'I'm having this' he cries. 'You said I could take anything I want. So I'm taking this'. . . It's the essence of a degenerating mind, periodically to lose all sense of continuous self, and therefore any regard for what others think of your lack of continuity." And Baxter's willingness to hope, to allow himself to be duped by Perowne into thinking he could join a clinical trial of RNA interference, is further proof of his cognitive failure.

Baxter is the worst caricature of someone with Huntington's disease.



**Saturday**  
Ian McEwan. Jonathan Cape, 2005. Pp 288. £17.99. ISBN 0-224-07299-4.

His calumny is blamed entirely on the length of the CAG repeat on his fourth chromosome, and his villainy is due only to his disease. McEwan sadly reinforces the stigma and stereotypes from which families with Huntington's disease suffer, and which make them hide both their inheritance and their destiny.

Statistics show that people with Huntington's disease are no more prone to violence or crime than anyone else in the population. Their bizarre and uncontrollable movements may be frightening, but they are no more dangerous than anybody else. One of us (Nancy Wexler) is at risk for Huntington's disease. Her mother taught high school biology; three of her uncles were professional jazz musicians; and her maternal grand-

father sold lingerie. All five died from Huntington's disease—but none were in any way violent.

As well as McEwan's pitiless stereotyping of Huntington's disease, the reader is also subjected to the most extraordinary personal and pharmacological name-dropping. Among many other individuals, even UK Prime Minister Tony Blair is made to put in an unflattering (and completely unnecessary) cameo appearance. The pharmacological name-dropping extends to dopamine, GABA, glutamic acid decarboxylase, choline acetyltransferase, and RNA interference that intersperse the text without the slightest indication of what the phrases mean. For most general readers such references will be unintelligible.

There is, though, an irony about this book. Although McEwan doesn't make it explicit, we only needed to exercise some amateur sleuthing skills to discover that Perowne's house is set in London's Fitzroy Square. In the corner of the square there is a statue of Francisco de Miranda. It is poignant that, so close to Perowne's house, there stands a memorial to the man who tried to deliver independence to the Venezuelan people; for it is thanks to these people's DNA that we have learned about the genetic basis of Huntington's disease—and the source of Baxter's fate.

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**Fed Up! Winning the War Against Childhood Obesity**  
Susan Okie. Joseph Henry Press, 2005. Pp 322. \$27.95. ISBN 0-309-09310-4.

## In brief

### Book Fighting back against fat

In *Fed Up*, physician-journalist Susan Okie diagnoses the multitude of factors that are driving the childhood obesity epidemic. Too much food and too little exercise, of course, but it's a bit more complicated than that.

In the USA, about 15% of children aged older than 6 years are obese, while another 15% are overweight. Given the societal and economic pressures at work to keep children wolfing junk food while planted in front of the television or computer, these children are shaping up to be the first generation in the nation's history with a life expectancy shorter than that of their parents. And while the USA leads the world in this particular race, the rest of the world is quickly catching up.

Okie lays out what seems like every current piece of research on this topic, from the role of genes and maternal weight during pregnancy in obesity to the effectiveness of school-based exercise programmes and the

feasibility of gastric bypass surgery for morbidly obese young people. She interviews leading researchers, clinicians, teachers, and community leaders. Most importantly, she spends time with kids at school, at home, and in the doctor's office, and relates how young people and their families are trying to develop healthier habits, often with encouraging success.

Given the difficulty grown-ups have keeping slim, it's daunting to imagine how kids can manage in a world where junk food is marketed so pervasively and, one could argue, insidiously. Vending machines sell chips, sweets, and sodas on the grounds of most US schools, and one in ten schools have "pouring rights", or an exclusivity contract, with beverage makers like Pepsi or Coke. The dual mission of the US Department of Agriculture's school lunch programme—to subsidise farmers and to nourish children—means offerings are so heavy on meat, potatoes, and dairy that one

student tells Okie she longs for salad every day.

Okie prescribes solutions to this "public health epidemic", but they're not simple. Families, schools, and communities must work together to encourage children to eat healthily and stay active, she says. Among her suggestions are working with farmers to bring fresh foods into schools; building environments where it's easy and safe for children to get around by bike or on foot; and limiting the time children spend watching television or playing on computers. Doctors have a role to play too, Okie says, which too often they ignore. One paediatrician tells Okie her colleagues may fear triggering eating disorders by addressing the issue of overweight with their young patients. As *Fed Up* shows, it will take a village—and much more—to help children grow up healthy in our media-saturated, over-fed world.

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