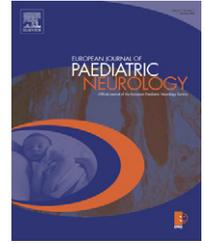


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## Original clinical historical contribution: Stuart Green Vignettes

### Stuart Green's Vignettes 11 and 12

#### 1. The voice is a clue

There is a disease called Batten's disease—a slowly progressive neurodegenerative disease of children—which causes visual problems which are progressive. The children become blind between the age of 5 and 8 and then they lose cognitive skills and eventually plateau leaving them in a state where their intellect is dulled, their speech is affected and they walk and move in a Parkinsonian-like way. The speech is stuttering and repetitive and when one sees a patient with this disease it is fairly distinctive.

I was visiting a school for the blind called Pondover, a wonderful school in an old mansion house in the heart of Shropshire. I had come as one of the visiting consultants. It is a big house with marvellous grounds and the children, all of whom were at least visually handicapped and some had other problems as well, were playing in the gardens. Aware of our presence a young girl of about 14 years, severely visually impaired for other reasons but quite able, came up to me and said "Hello, who are you?" I told her who I was and she asked me why I was there and I said "I have come to visit Alison and John" (the children with Batten's) "Oh, we all know Alison and John" she said "they've got the same thing." "What do you mean?" I asked. "Well we call them the 'mumbles' because when they speak they go 'mumble, mumble, mumble'. It is very funny the way they speak." Here was a blind girl, who knew nothing about Batten's disease at all, nothing about medicine, but who had identified that two of her classmates had the same condition because of the unique way they spoke.

*Comments*

What wonderful insight!

#### 2. The eyes (or eyelids)

Even in these days of modern clinical scanning by chemistry and specialist investigations, close clinical observation still has a large part to play.

I was asked to see a young boy about the age of 11 or 12 years who appeared to have gradually deteriorated in general health over a period of 3 months. He had at first some

difficulty with eating, lost weight, become tired and lethargic, complained of pains and aches and gradually become weaker. He was almost wasting away, he wasn't able to go to school, he had difficulty in swallowing, his speech became limited. Extensive investigations had failed to show any obvious cause in terms of a brain tumour, an infective process or metabolic process. It was felt he had some insidious brain disease which had not shown up on the scans, which had caused him to waste away and lapse into a semi-conscious phase.

I was asked to see him at another hospital. His parents were naturally very anxious about him. They told me he had been a bright boy; he had recently started secondary school and appeared to be coping quite well. There had been one or two problems at school but nothing they thought was very significant. He had not had any infections or any injuries prior to the illness starting. When I saw him he was lying on bed, he looked pale and thin, he was fed by a naso-gastric tube. He was able to move about in the bed and roll from one side to the other and when one spoke to him there was only the minimal recognition that he could hear anything at all in that he responded with almost a grunt. It was difficult to get him to obey any command at all and he lay almost curled up in a ball and mute. Apart from him being not very responsive I found very few abnormal findings apart from his wasting, except one. When I tried to examine his eyes with an ophthalmoscope, I noted two things. First of all his eyelids were fluttering to some extent and when I tried to open his eyelids to look in his eyes his eyelids flexibly closed. Now most children in a coma or a semi-coma or stupor, when stimulated or provoked, will either not open their eyes or open their eyes partially if they are roused. What they won't do (there is no mechanism known for this) is *close their eyes more tightly*. This made me very suspicious that this child did not have the significant brain disease he was thought to have by his attendants. I brought him over to our own Children's Hospital and saw him together with a colleague of mine, a very experienced neuropsychiatrist, who agreed with me.

*Comments*

It often requires considerable neurological skills to be sure that symptoms and signs are impossible—impossible for any of the central nervous system diseases that we paediatric neurologists deal with. Though Stuart is pretty sure of the

diagnosis, he calls in a child neuropsychiatrist experienced in this kind of problem<sup>1,2</sup>. The difficulty then is to give a positive and encouraging explanation to the family, but a problem shared is a problem halved.

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