

UNCOVERING INCONTINENCE

Several of this month's papers look at uncommon conditions from the point of view of families. An editorial by parents of a child with precocious puberty reminds us that clinical and scientific uncertainty can be hard to bear. But even when there is little doubt about diagnosis and management, outcomes as seen by the sufferer may differ from that assumed by their clinicians.

A team from Great Ormond Street Hospital in London, UK report a 7–17 year follow up of 30 children with Hirschprung's disease divided according to whether their aganglionosis as total colonic (TCA) or rectosigmoid (RSA). Surgeons in the team studied continence by history, examination, and anal endosonography while a psychologist did the same using a semi-structured interview. The latter also investigated the children's competencies, behaviour, emotional state and self esteem.

Incontinence was depressingly common. The authors did not find an association with psychosocial maladjustment but caution that their numbers are small. The surgical investigator was more likely than the psychologist to conclude that children were continent—a methodological lesson for those conducting follow up studies, even when looking solely at a physical symptom. Should we be surprised that a psychologist's questions might probe deeper than a surgeon's finger? We look forward to receiving more such cross-specialty studies.

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THALASSAEMIA AND THE LIVER—10 YEARS ON

Investigators from Hong Kong performed liver biopsies on 73 patients with thalassaemia major who had received chelation for a median of 7.7 years and transfusions for a median of 10.6 years. Nearly half had received bone marrow transplants.

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All had evidence of haemosiderosis, its extent correlating closely with serum ferritin but not with the duration of chelation or transfusions. Hepatic fibrosis, likewise, was associated with a high ferritin level (and with hepatitis C infection). The authors found no diminution in severe grades of haemosiderosis when compared with studies conducted elsewhere 10 years previously. They attribute this to poor local compliance with chelation therapy. They advise using serum ferritin to monitor children who have not yet developed serious iron overload. A high ferritin or evidence of hepatitis C infection requires liver biopsy to evaluate the degree of fibrosis and to measure liver iron content.

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BAYES' THEOREM AND BRUISES

Front line paediatricians may be drawn to a diagnosis of child abuse, in part by the extent and pattern of bruises on a small child. Improving the accuracy of diagnosis is obviously important. A group in Wales have suggested previously that Bayes' Theorem might help.¹ For the few readers unfamiliar with this, our statistical adviser, Michael Healy comments that it is a formula for updating one's probability beliefs in the light of receiving a piece of evidence. This sounds to me as presumably what good clinicians should do instinctively. Professor Healy adds that its most common medical application is deriving a predictive value from prevalence following a test result, using the sensitivity and specificity of the test.

The investigators have now looked at site, shape, and dimension, subsequently calculating a score by logistic regression analysis. They suggest that, in practice, a clinician would first estimate the prior probability of abuse given the clinical situation; he or she would then calculate a bruise score and use a sensitivity and specificity table (table 3 of the paper) to estimate the posterior odds of abuse. Testing the score on children seen by the investigators suggests it is overoptimistic and they intend to continue to test in on more children to define how robust it might prove to be.

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LITTLE ORPHAN ANECDOTE

A few years ago I received a request from a Norwegian paediatrician to provide an opinion on his patient. She had heard that I was a world expert on the condition. Despite my not being a world expert on anything in particular, I did my best to assist. The reason for his misapprehension was that my name had been quoted by a parents' support group which had heard that I had 2 probables and 1 possible under my care.

Looking at some of my academic colleagues I had often thought that some must have been born experts while others acquired the title. Now here was I having it thrust upon me. I prowled around Medline looking for the real thing without success. More recently I returned to the fray by asking our commissioning editor, Nick Mann, to obtain an editorial on the subject: hence, this month you can read the words of Drs Carter and Metcalfe from NIH on the mysterious condition of systemic mastocytosis. Now you can all be your own world expert.

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AND FINALLY . . .

Now that ADC has stopped using envelopes and stamps, we have encountered the new sport of literary automatic responses to our emails. Here's one from Prof Virginia Sybert, a Seattle dermatologist, to whom we sent a computer generated letter requesting she referee a paper:

*When it comes to messages electrical
My response is somewhat eclectic
I may answer ways rapid
But if my brain's vapid
You may not hear from me this week or nextical.*

The great McGonigall, tragedian and poet had best look to his laurels.

¹ Kemp AM, Kemp KW, Evans R, et al. Diagnosis of physical abuse using Bayes' theorem. *Child Abuse Rev* 1998;7:178–88.