Dear Sir,

'Vett Syndrome: some behavioural aspects and an overview'

Having made a particular study of Rett Syndrome since 1982 and with experience now of several hundred cases we wish to record complete disagreement with the unsupported opinions of Drs Elian and Rudolf regarding the nature of the characteristic repetitive hand movements.

Following careful evaluations based on continuous non-invasive monitoring of breathing and limb movements, blood gases and the electroencephalogram, we have described the course of events throughout life and suggested that the age related movement and respiratory abnormalities are essentially involuntary, possibly part of a 'package' of extrapyramidal accompaniments to alerting of the girl (1, 2, 3). There is no evidence that the girls with their specific incapacity for voluntary movement, are able to plan and direct these complex and entirely predictable movements; however there are indications that the girls may become distressed and exhausted by them. These rhythmic movement patterns may be observed in some girls long before regression begins. They have also been recorded immediately upon waking while there is no other person in view.

Elian and Rudolf's use of the term dementia in Rett Syndrome is ill advised since women over forty years of age appear no more disabled mentally than they were following regression. While such a devastating disorder must carry inevitable sequelae for the ageing brain no evidence has been found of primary dementia. The nature and timing of the regression phenomenon itself are suggestive of a neurodevelopmental phenomenon linked to maturation of neural control (3, 4, 5).

Yours sincerely,

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References
Dear Sir,

Drs Kerr and Southall's letter, written in 1991, critical of our paper which appeared in 1989 shows a surprising delay, the reason for which escapes us. That distinguished research team “with experience now of several hundred cases”, who “made a particular study of Rett syndrome since 1982”— in the UK surely did not for so long overlook an overview of Rett syndrome published in England by a fellow research team? Our studies are based on 16 patients, a modest number compared with that of Dr Kerr et al., whose speed of collecting Rett girls is admirable: in April 1991 they wrote to us—via Acta Neurol Scand—that they had seen “over fifty” patients with Rett syndrome; in their letter of May 1991—via Behavioural Neurology—this number rose steeply to a staggering “several hundred cases”.

It is not unexpected that a newly identified disease of so far unknown cause will not enjoy universal consensus on all its aspects; this explains the many teams working on Rett syndrome all over the world. Therefore, we are not surprised to note that Drs Kerr and Southall are in “complete disagreement” with us. They are entitled to their particular opinion supported by their methods of study; and of course we are entitled to ours, well supported by the results of our study and by Professor Rett’s opinion.

It seems that the authors of the letter have misread or misunderstood several points in our present paper, as they misunderstood the statement on low CO₂ in our paper in Acta Neurol Scand, which so recently they have disagreed with, only for it to emerge that basically our opinions are similar.

For example, the letter writers carefully point out that the abnormal hand movements or respiration were also recorded “while there was no other person in view”; the spirit of this sentence seems to us to accord with our statement that the hand movements “are seen especially when the girls are left alone”.

Kerr and Southall argue that “respiratory abnormalities are essentially involuntary”, disregarding the well known fact that breathing in man (girls included) is regulated by two anatomically distinct but functionally integrated respiratory control systems, one of which mainly operates during wakefulness—the so-called behavioural or voluntary system—while the other is active during NREM sleep, the so-called automatic or metabolic system. The regulation of ventilation by the behavioural system is in use while the subject talks, experiences anxiety or fear or holds her breath. It is this behavioural or voluntary breathing mechanism we are talking about.
and of course we did not expect, nor found evidence that these girls "are able to plan and direct . . ."); clearly that is a misunderstanding of our views. Kerr and Southall have found that the hand movements are "entirely predictable"; we have found that, while stereo typical for a given girl, they seem to be rather unpredictable as far as timing or speed is concerned.

Finally our view on dementia stands, as it satisfies various definitions including that of Mayer Gross, quoted in our paper.* The fact that "women over forty years of age appear no more disabled mentally than they were following regression" is irrelevant regarding the alleged improper or "ill advised" use of the word "dementia". We did, however not make it sufficiently clear that the regression stops and the dementia ceases to progress after a time. *Nostra culpa, solely on this account.

Yours sincerely,
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* Dementia refers to a global deterioration of mental functioning in its intellectual, emotional and cognitive aspects; intellectual decline is the central feature.

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