aposition of the cords can occur normally during quiet
sleep in young lambs. Briefly, this ‘braking’ of expiratory
airflow by the larynx generates positive subglottic
pressure of up to 10 cm water and is an important
mechanism in preventing lung collapse in the young until
a noncollapsible chest wall is established. If this expiratory
laryngeal pressure is prevented, such as by a tracheotomy
or endotracheal intubation, respiratory frequency falls
and respiratory drive is greatly diminished especially
during quiet sleep. Equally, it can be reasoned, if an
overdistensible trachea exists or develops, respiratory
drive could diminish, and failure or ‘unexplained death’
occur during respiratory challenge—such as with an
apparently trivial infection or upper airways obstruction
during sleep. That is to say, positive expiratory pressure
could not be developed if the upper airways simply
dilated with glottic adduction.

What subglottic pressures are generated in respiratory
disease in the young appears to be uncertain. From our
studies in early postnatal life we would expect tracheal
distension during the expiratory cycle (rather than
obstruction) in quiet sleep but such airway collapse could
occur in rapid eye movement sleep when upper airway
expiratory resistance is slight.

Thus a hypothesis to be tested could be an association
between vocal cord necrosis, indicative of chronic
laryngeal adduction in expiration, and a distended
trachea in these cases. The information from this
comprehensive series of necropsies could provide valuable
evidence which could point the way to a functional
evaluation in such infants possibly averting later morbidity
and mortality. Surely a morbid anatomist’s dream!

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Gluten intolerance, gluten
enteropathy, and coeliac disease

Professor Emery and Dr Wailoo comment:

We were very well aware of most of the points raised by
Paul Johnson but did not wish to speculate too far beyond
our evidence.

The possibility of spasmodic glottic closure being a
major factor in unexpected death is one that we have been
studying for many years but unfortunately, as with so
many things related to cot death, these vocal cord lesions
also occur in children who die in hospital from apparently
clinically justifiable disease. It would be possible, and

nice, to do the anatomical correlations that Johnson
suggests but the nonspecificity of the vocal cord lesions
would be likely to lead to an inconclusive result.

Our own interest has progressed from the general
statistical conclusions to the possibility that there is a
particular area of tracheomalacia at the carina which
enables the trachea to ‘block off’ at this point and that
infection is either absent or present at the laryngeal conus.

It would be of interest to us if Paul Johnson could give
his findings regarding the presence or absence of laryngeal
lesions indicating spasm in his lambs!

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Dr Dodge comments:

I am grateful for Professor Salazar de Sousa's comments. I had intended my paper to be provocative.

In my own practice, I follow the ESPGAN protocol and submit patients to three biopsies. However, this procedure is followed by only two-thirds of the members of ESPGAN and I imagine that only a few nonspecialist general paediatricians in this country routinely perform a gluten challenge, followed by a biopsy. Moreover, the challenge procedure varies considerably from putting the patient on a free diet to daily administration of a prescribed large amount of gluten. The postchallenge biopsy is essential if a diagnosis of persistent gluten enteropathy is to be made. I want to encourage paediatricians who are daunted by the present protocol to give a gluten challenge to patients who have benefited clinically and, if known, histologically from a gluten-free diet.

Most of my patients in Cardiff seem to adhere very well to their dietary regimen, and it is exceptional to find one in whom the mucosa has not recovered when a second biopsy is performed. However, simply knowing that the mucosa is still abnormal would not ensure future patient compliance, and I believe that our good results can be attributed to the continuing advice and encouragement given to the parents by our dietician. The primary purpose of my paper was not to challenge the usefulness of the ESPGAN criteria (and I agree that we have nothing better at the moment), but to suggest that until we have a satisfactory definition of coeliac disease that is accepted both by physicians and paediatricians, we could improve the clarity of our communications by using the terms 'gluten intolerance' and 'gluten enteropathy' to describe clinical symptoms and histological appearances. When Professor Salazar de Sousa tells me that a patient has coeliac disease I understand him perfectly because we both use the same diagnostic criteria, but it seems more sensible to use accurate descriptive terminology when the investigative process is incomplete, or when the criteria are not agreed.

References


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Dissolution of bilateral cystine calculi by penicillamine

Sir,

I read with interest the paper by Ruysch van Dugteren and Wiggelinckhuizen.1 I have obtained similar results with alpha-mercaptopropionyl glycine. Cystine-lysine-ornithine-argininuria was diagnosed in two siblings—one with bilateral staghorn calculi—by using high-tension electrophoresis. Calculi were dissolved by means of alpha-mercaptopropionyl glycine in the child with bilateral calculi (Figs 1 and 2), while prevention, without presence of calculi, was achieved with the same drug in the other child. These results show that the drug is useful

Fig. 1 Patient aged 5 years: (x-rays of abdomen). Bilateral renal calculosis (arrows).