et al, recently reported 14 cases which they documented in this manner.1

Within a period of six months we have seen three babies with ALTE from three unrelated families. All three had been discovered limp, cyanotic, and apparently lifeless during their afternoon nap. Petechial haemorrhages were found on the face and neck of two of the babies on admission. No further episodes occurred during observation in hospital or the follow up period.

These would probably have remained unexplained unrelated cases were it not that when the third case was brought to the casualty department by the family doctor he was accompanied by the babysitter who had discovered the baby. She was recognised by one of the nursing staff as a regular attendant at the casualty department with minor wounds that were suspected of being caused by autamutilation and she had made several allegations of being attacked or raped.

In the ensuing discations it came to light that this woman was the babysitter who had also been involved in the first two cases. She has since been investigated by the police. However, there are strong suspicions that, if imposed upper airway obstruction of the babies by her, there has been insufficient evidence to bring her to trial.

CVS as described by Southall et al2 would have failed in these cases, because it is usually only the parents or very close relatives who are allowed to be continually present with the baby in hospital. Very thorough history taking especially concerning the surrounding circumstances remains extremely important in investigating every case of ALTE especially if imposed upper airway obstruction is suspected.

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1 Samuels MP, Mc Claughlin W, Jacobson RR,
2 Southall DP, Stubbens VA, Rees SV, Lang MH,
Wynne CH. Community paediatric cases induced by smothering: two cases identified by covert video surveillance BMJ 1987;294:1637-41.

Anal abnormalities in childhood myotonic dystrophy

SIR.—The paper by Reardon and colleagues on anal abnormal signs seen in myotonic dystrophy is worrying on several counts.1 The physical signs are described inadequately which makes interpretation of the paper difficult. The Royal College of Physicians report,1 although not referenced, is not heeded in the need for a consensus on definition and method of examination. Thus ‘a reflex dilatation of the anus was observed on parturic the buttocks’. How was the child examined, for how long, and was this a dynamic sign and what degree of dilation was observed? It is noteworthy that only one child in six was said to demonstrate this sign but all had anal laxity.

The illustration shows a degree of anal laxity we have never seen, is this laxity or dilation? Was this degree of gaping achieved on squeezing the buttocks? Was the child constipated and demonstrating the ‘visibly relaxed anus’ of Clayden?2 This child was 15 years old and had had a lifetime of soiling, what had earlier examinations revealed and to what treatments and manoeuvres had she been subjected?

Single physical signs are rarely diagnostic and in making a diagnosis of child sexual abuse the jigsaw must be carefully constructed.3 To begin an investigation on the basis of a child with known bowel function and anal laxity is clearly problematic but in the context of a girl who is alleging abuse it is, in child protection terms, quite proper. Advice to doctors stated ‘It is important to take what the child says very seriously, and to spend time listening to what the child has to say.’ It is also evident that children with special needs are at risk of abuse and to recognise abuse in children with communication problems requires particular skill.4 Children do need protection from the trauma of wrongful diagnosis but similarly physicians have until recently failed their patients by their inability to recognise maltreatment.

Finally, what proportion of children with myotonic dystrophy have bowel disorders or an abnormal anus on examination? What proportion have no abnormalities? The association of two conditions does occur.

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Drs Reardon, Hughes, and Green and Professor Harper comment:
The comments of Drs Wynne and Hobbs reflect the difficulties with which clinicians are obliged to grapple in trying either to confirm or refute a suspected diagnosis of child sexual abuse. Foremost among these is the non-specificity of single clinical signs. However, in many instances, abnormalities of anal physiology do contribute significantly to decisions with far reaching effects for patients and families. It is precisely because we absolutely agree with Drs Wynne and Hobbs that children need protection from the trauma of ‘wrongful diagnosis’ and because of the central role which anal abnormalities and interpretation thereof often has in reaching diagnoses of sexual abuse that we felt prompted to submit our report.
The report did not pretend to be a scientific treatise of anal sphincter dysfunction in myotonic dystrophy and the role of non-specific clinical observations is to make colleagues aware of added potential pitfalls which attend the diagnosis of child sexual abuse in myotonic dystrophy. We feel that shared clinical experiences such as ours are of value to clinicians in avoiding inappropriate diagnoses of child sexual abuse.

Prostacyclin concentrations in haemolytic uraemic syndrome after acute shigellosis

SIR,—Although the epidemiology of certain forms of the haemolytic uraemic syndrome (HUS) has become clearer in recent times, pathogenic details remain largely unresolved. The role, if any, of prostacyclin (PGI2) in the disease associa ted forms of HUS (D+HUS) cannot be deduced from previous reports and the study by Alam et al on patients with shigella induced HUS adds to the confusion.1 In the following we acknowledge that past European and American studies the aetiology of D+HUS is likely to be verocytotoxin producing Escherichia coli, which although similar is not homologous with shigella induced HUS.

Prostacyclin is probably not a circulating hormone in man: its plasma half life is brief and intact molecules cannot be measured directly in clinical practice. Thus the term 'prostacyclin concentration' is at best an extrapolation. Two main approaches have been used to identify abnormalities of PGI2 metabolism in disease states. The more direct of these is to measure excretions of metabolically stable products such as 6-keto-PGF-1α in biological fluids. The commonly used radioimmunoassay kits give plasma estimates which exceed those of the more specific gas chromatography and mass spectrometry methods. Blood sampling in itself induces artifacts, and although the urinary excretion of degradation products may be more consistent in health, this is unlikely to be a reliable method when renal function is rapidly changing. Both Stuart et al5 and Turi et al6 found plasma concentrations of prostacyclin metabolite to be increased in D+HUS at onset.

A second approach has been to determine whether plasma or serum stimulates generation of prostacyclin by endothelial tissue in vitro. Prostacyclin release into the supernatant medium is measured indirectly by bioassay or as 6-keto-PGF-1α. Published results are conflicting: Schlegel et al7 and Levin1 found that prostacyclin generation was promoted by plasma, whereas Turi et al6 and Siegler et al8 found reduced concentrations. Methodological differences may explain these inconsistent findings, but abnormalities of plasma stimulating factors or inhibitors have in any case not been shown to reflect disturbances of prostacyclin metabolism.

Unfortunately Alam et al did not make clear which method they used. The result section and abstract refer to plasma concentrations of 6-keto-PGF-1α suggesting direct measurement. However, their method refers to radioimmunoassay for 'prostacyclin', but also to generation of 6-keto-PGF-1α by rabbit aortic rings in response to patients’ plasma.

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1 Alam AN, Abdul MA, Wadah MA, et al. Prosta- cyclin concentration in serum from patients sharing the non-specific clinical observations is to make colleagues aware of added potential pitfalls which attend the diagnosis of child sexual abuse in myotonic dystrophy. We feel that shared clinical experiences such as ours are of value to clinicians in avoiding inappropriate diagnoses of child sexual abuse.

Archives of Disease in Childhood 1992;67:1412

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