

- According to Gnanaraj *et al*,¹ none of the nine fatalities with putative crush head injuries in Group 2 had retinoschisis or folds, but peer-reviewed publication of these fundal findings has not occurred in any of the purported child abuse (shaking) fatalities from that 20-year-old data set.⁴
- The authors misstate that crush injuries to the head are the 10th most common cause of injury in children under the age of 9 years. The cited reference lists crush injury as the 10th most common cause of injury in children under the age of 9 years (ICD-9 codes 925–929).⁵ This specifically excludes intracranial injury (ICD-9 codes 850.0–854.1).⁶
- Gnanaraj *et al*¹ claim that the perimacular retinal folds observed in the child from the evidence-based case report by Lantz *et al*⁷ were a bit atypical, more angulated at the apex; however, the cited reference does not discuss this subjective nuance nor was apical angulation of the retinal folds described in the case report or evident from the accompanying images.⁸
- Gnanaraj *et al*¹ state that retinal haemorrhages are well recognized although uncommon in accidental major head trauma citing a 1992 study.⁹ This article has been previously identified as exhibiting selection bias based on the relative minor head trauma sustained in the accident group.⁷ Similar systematic error is obvious in the selection of patients (Group 1) by Gnanaraj *et al*¹ when compared to previously published studies characterizing the morbidity and mortality of head injuries associated with falling televisions.^{10–12} Remarkably, four of the nine children (44.4%) with accidental head injuries in Group 2 by Gnanaraj *et al*¹ had retinal haemorrhages.

All too often, the human tendency is to embrace repetitious assertions that reinforce authoritative opinions, but trivialize or reject new findings that do not support entrenched beliefs.¹³ The perpetuated claim that retinoschisis and perimacular retinal folds in children are created by vitreoretinal traction during violent shaking resides in faith not in science.

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Sir, Response to Drs Lantz and Stanton

We read with interest the comments made by Drs Lantz and Stanton and thank them for their interest in our work.

- (1) Retinosis is a term, which means splitting of the retina. This is an objective finding readily identified on standard histological preparations of the retina. Although we certainly agree with Drs Lantz and Stanton that the well-known and documented association with abusive head trauma was first described in 1986, the actual identification of the histological finding could have been made by histological examination for many years before. The ocular findings were assessed simply by having an ophthalmic pathologist examine the eyes and record what was observed. Likewise, the ophthalmic findings in the nine fatalities, where there were no folds or retinoschisis, do not require peer review any more than any clinical findings in the literature. The slides were prepared according to the protocol¹ and review. We reported the result and it was peer reviewed for publication.
- (2) Although clinically insignificant, we thank Drs Lantz and Stanton for indicating our error in citing crush injuries as the 9th most common cause for injury in

children below nine years of age; it should have been the 10th cause. Intracranial injuries excluding skull fractures are the 6th and skull fractures excluding vault and base fractures are the 12th most common causes.

- (3) Dr Lantz first presented the case in 2002 (Lantz P, Sinal S. Perimacular retinal folds in non-abusive head trauma. *Fourth National Conference on Shaken Baby Syndrome*. Salt Lake City, UT, 2002). One of our authors (AVL) had the privilege of attending this presentation after which Dr Lantz kindly sent him images from the original histological preparations including images, which did not appear in the published description.²
- (4) There is copious evidence to support the observation that retinal haemorrhage is uncommon in accidental head trauma. This evidence is well beyond the scope of this reply and the reader is referred elsewhere.^{3,4}
- (5) Three of the four children with retinal haemorrhage had few, small haemorrhages largely confined to the posterior pole. The fourth child also had microscopic (ie, not clinically detectable) haemorrhage and a few that were visible on gross inspection at the ora serrata. That child also had significant neck injuries consistent with atlanto-occipital disruption. It would be improper to suggest that this high percentage (4/9, 44.4%) should be used in any way as a reflection of the potential rate of haemorrhages in other types of accidental head trauma. The rate is entirely inconsistent with a literature that examines tens of thousands of eyes in victims of non-abusive injury. Although the high percentage may simply be a reflection of small sample size, the higher prevalence may simply reflect, if confirmed on a larger sample size, the severe fatal crush injury mechanism (head out of window when car rolled, car rolled over head, unrestrained back seat passenger, and pedestrian struck by car), and even then it is remarkable how mild the retinopathy is and how it differs from that seen in two-thirds of shaken babies.⁵

Although we certainly value the examination of new data and outlier reports,⁶ we regret that Drs Lantz and Stanton reject the large body of medical literature, which not only supports the current understanding of the pathophysiology of retinal haemorrhage in abusive head trauma but also weighs very heavily against the paucity of evidence to the contrary.

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Sir, A novel peripherin/RDS mutation resulting in a retinal dystrophy with phenotypic variation

Peripherin/RDS is a structural transmembrane glycoprotein that contributes to the formation and stabilisation of rod and cone photoreceptor outer segment discs. Mutations in the peripherin/RDS gene can result in generalised retinal dystrophies or macular dystrophies^{1,2} and are known to cause variable manifestations within families. We describe a novel mutation in exon 2 of the peripherin/RDS gene resulting in a three amino-acid deletion and causing a variable retinal phenotype within a three-generation family.

Case reports

Case 1

A 30-year-old man presented with a history of nyctalopia and reduced-peripheral visual fields. His best-corrected visual acuities were 6/6 in each eye. Anterior segments were normal on slit-lamp examination. Fundus examination revealed punctate hyperpigmentation in the peripheral retina (Figure 1). Colour vision was normal. Electroretinograms showed a reduction in light-adapted