

LETTERS TO THE EDITOR

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Life expectancy in cerebral palsy

EDITOR,—The recent study by Hutton, Colver, and Mackie¹ is in some respects a useful addition to our knowledge of survival in cerebral palsy. Unfortunately there are substantial problems with the paper; we note three of them below.

1. In figure 1A it appears that in the most seriously affected group, who had a Lifestyle Assessment Score (LAS) $\geq 70\%$, there is 100% survival to age 9. This scarcely seems plausible when, as the graph indicates, 20% of these survivors die in the next 9 years. The explanation is that the most severely disabled children, with LAS 70% or more, have to survive to age 5 to be assessed by LAS. Thus the severely disabled children who die before 5 have no LAS, and are excluded (actually, it appears from the graph that some children are evaluated even later than age 5).

The resulting bias could have serious consequences. For example, in a lawsuit involving a neurologically devastated 2 year old child a plaintiff may cite Hutton *et al* to argue for 100% survival over the next seven years.

2. Hutton *et al*'s results show that, as is well known, low IQ and/or poor mobility correlate with reduced life expectancy. In his commentary, Dr Rosenbloom usefully asks whether extreme immobility or mental impairment would give an even greater reduction. The answer is clearly yes, as indicated by our own work^{2,3} and is also acknowledged by other workers in the area.^{4,5} Indeed it must be so because an extensive literature shows a much shortened survival in the persistent vegetative state,⁶ which may be regarded as the extreme case of disability.

In neither HCM nor in Dr Hutton's previous study of the Merseyside area⁷ is this possibility mentioned. As a result, the latter study has frequently lead plaintiffs to overestimate survival of children with the most severe disabilities.

3. In their table 5, Hutton *et al* state that in our California study² the odds ratios for various hazards were lower than in several other studies. For example, in table 2 of our article we gave a hazard ratio of 3.8 for two year olds who were tube fed, compared with those who were not. Hutton *et al* then proceeded to

speculate at some length on what accounts for the transatlantic difference.

The real reason is simple: the California database has many more variables other databases, so the marginal effect of any one of them—that is, when the others are held constant—is smaller. For example, our table 1 showed that if no other factors are taken into account the hazard ratio for tube feeding (compared with children who could self feed) was 23.6—a much larger ratio than the above 3.8, and in fact about as large as any in Hutton *et al*'s table 5. In addition, the definitions of mobility etc in the various studies are very different.

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Reply

We thank David Strauss for his interest in our work but he fails to substantiate his claims that there are "substantial problems" with it.

First, we wish to correct an error in our article on page 470, column 2, line 11: "dying before" should read "surviving until".

With regard to Strauss's remarks on LAS, both the abstract and the results section include the phrase "survived to age 5", so Strauss has not explained our result, but merely repeated this information. Even the brief precis of the LAS in the paper makes it clear that it would be difficult, if not impossible, as well as unwise, to attempt to complete it for a 2 year old. That someone might misquote our work is true, but not our responsibility.

With regard to mobility and mental ability, we have reported exactly what is measured, and have referenced other work which includes measures different from ours. It appears that Strauss wishes we had speculated about information we do not have. Note that IQ was constructed to have a mean of 100, and standard deviation of 15. On this scale, fewer than 4 in 10 000 people would have an IQ less than 50, our definition of severe cognitive disability. Fewer than 5 in 10 million people would have an IQ of less than 20, the number mentioned by Dr Rosenbloom.

It seems obvious that persistent vegetative state or indeed leukaemia or need for ventilation twice a year are clearly additional, and largely independent risk factors for death which any court would take in to account.

With regard to hazard or odds ratios, our discussion does comment on multivariate versus univariate models. Whether univariate or multivariate, one still has a relative statistic, so that any lack of similarity in the baseline categories will be relevant. Strauss claims that "the real reason is simpl(y)" the

difference between multivariate and univariate analyses: he gives hazard ratios for tube feeding, and states that the univariate ratio of 23.6 is "about as large as any in Hutton *et al*'s table 5". However, Strauss fails to quote a factor—hand use—which, unlike tube feeding, can be easily compared with our work, and that of South East Thames. In correspondence with Hutton, Strauss stated "Re hand use: our multivariate OR was 1.52 and our univariate OR was 5.69": a value which is substantially less than 23.6, and than our results. Further, Strauss states that this lower ratio is for a more disabled group: "But our definitions are very different from yours. Our 'bad' group is 'no functional use of hand'...while your 'severe manual disability' group is much more inclusive'...". Thus the "reason" Strauss gives fails to explain the difference in results for manual disability.

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Evolving practice

EDITOR,—I am aware that a respected scientific journal does not normally indulge in political issues but you seem to be setting a precedent by "Reflecting on Redfern".

As a registrar at the Alder Hey Hospital in the 1980s, I was one of those taking consent for post mortem from parents of children dying in the cardiac unit. It was one of the most difficult jobs I have ever had to undertake. It was done, not to provide specimens for museums, but to provide parents with as much knowledge as possible about why their child died. It was regarded as the parents' right to have this information, and that was the spirit in which consent was obtained.

It is true that details of the procedure were not volunteered but neither were they withheld if requested, which was hardly ever. As many people have commented, it was not the intention to deceive but to avoid distress. The lack of probing by parents only seemed to confirm their wish not to know. I believe I undertook this task with honesty and integrity. I feel no shame in my actions and have no wish to offer an apology.

Professor Hall was correct to say we should all be looking at what we do now, for which we shall be castigated in the future. Inevitably something will emerge but does this mean we are all currently acting in a paternalistic, arrogant, callous fashion. I do not think so.

If, in the 1980s, I had been required to gain specific permission for organ retention I could have accepted that as part of my job. However the system and parents did not request that I did. Why is it necessary to effect this change in practice in such an agonising fashion? The answer is in our malevolent British media, who are not content with evolving practice but need scapegoats and whipping boys.

We need as a profession to respond to changing expectations of society, but must we do so in such a self flagellating manner?

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Fertility preservation in children—scientific advances, research dilemmas, and ethics of consent

EDITOR.—The two publications on fertility preservation for children^{1,2} raise important issues but several issues need to be clarified.

Specifically, intracytoplasmic sperm injection (ICSI) is not a method to reverse male infertility in whatever circumstance. ICSI provides an effective solution to severe male infertility problem but offspring and partner issues need to be considered carefully.^{3–6} The suitability of pre-pubertal testicular tissue is questioned since technology surrounding cryopreservation and in vitro manipulation of prepubertal testicular tissue is stated as being “entirely experimental”. This is also true of adult testicular tissue which may similarly have immature germ cells.

The question of peripubertal boys and the use of rectal electrostimulation raises seriously important issues about the pain and psychological effect this procedure as a “first sexual experience” could have on the patients future sexual development and outlook.^{7,8} The procedure needs to be performed under anaesthesia. Any suggestion that this approach could be tried on peripubertal patients would be ill advised since aiming to obtain an ejaculate necessarily signifies post pubertal status and one has to be certain this level of maturity has been attained. This technique could be open to abuse, for in strict cultures where masturbation is forbidden, a parent could ask and consent to this procedure in post pubertal boys leading to a conflict in the requirement of an “autonomous consent”. Sperm storage under forced conditions will most likely be illegal, with possibilities of assault charges to the person taking the sperm sample. There remains also the probability of having mature sperm even if the patient has not yet reached the Human Fertilisation and Embryology Authority (HFEA) stated Tanner stage II maturity level. Such a situation would present a legally awkward sperm storage scenario with apparently no regulatory guidance.⁹

With respect to the statement that “fertility preservation procedures for children are experimental”, it is worth stating that the whole field of human assisted reproductive technology (ART), ranging from cryopreservation of sperm, oocyte, embryo, blastocysts, to the use of IVF, ICSI, and PGD has never undergone classical clinical trial evaluation. In this respect ART continues to be experimental, which is partly why the field is uniquely regulated by statute under the HFEA. There are advantages in bringing children’s gonadal tissue under regulation since in the eventual use for procreation the HFEA’s permission will be needed.

The second paper deals with the complex issue relating to the ethics of consent for gamete storage and experimentation. The title is broadly incorrect since children’s gonadal tissue do not contain “gametes”. Where gametes are contained we have adopted the term adolescents¹⁰ to help achieve consistency with reproductive biology, development, and regulatory framework which strictly requires consent under the HFEAct 1990.¹¹ The appeal for “intervention to preserve fertility to have sound basis for evidence and moral provenance” is reasonably well understood by

all practitioners but in the report² seems to signal “why things should not be done”. It should be remembered that the whole field of ART continues to be practised on its experimental origins worldwide reasonably sensibly and sensitively. Most of the consent models referred to in the text are the US style “assent type consent” which deserve acknowledgement.^{12,13}

The statement that “the HFEA recommendations were understandably not designed with children or cancer patients in mind” is not entirely correct in relation to cancer patients where the HFEA displayed enormous sensitivity and fairness. Following our representation on behalf of cancer patients in the year the HFEAct came into force, the HFEA responded by issuing a special direction to allow extension beyond the restrictive 10 year storage time for sperm, after considering the implication especially for adolescent cancer patients.

In the long term, efforts should be made to find ways of bringing childrens gonadal tissue under statutory HFEA regulation.^{14,15} This can be done by persuading Parliament through professional or patient representation to enlarge the regulatory remit of the HFEA, to help enhance patients interest, and in achieving consistent policies. Furthermore, it should be borne in mind that the UK legal landscape has changed significantly with the Human Rights Act 1998,¹⁶ favouring patients determination and sensibly engaging this Act may become a useful option.

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Life threatening airway obstruction in a newborn following ingestion of a sterilising tablet

EDITOR.—Sterilising tablets have been marketed and widely used to sterilise infant feeding bottles for the past 25 years.¹ Accidental ingestion commonly results in mild gastrointestinal irritation.² Severe airway obstruction has been reported in an older infant.³ Recently a 4 day old infant presented to our hospital with features of acute upper airway obstruction following accidental ingestion of sterilising tablets. The mother had found her struggling to breathe, with profuse secretions and smelling of sterilising tablets. She required prompt endotracheal intubation and mechanical ventilation for 48 hours. Massive swelling of the epiglottis, tongue, and arytenoids was noted at intubation. Severe stomatitis and oral ulcerations followed, requiring nasogastric feeding for two weeks. In addition to ventilation, treatment involved airway toileting to clear secretions, a short course of corticosteroids, and nutritional support.² Upper gastrointestinal contrast studies excluded oesophageal stricture. The tablet was given to her by her 4 year old sibling, who was very keen to share babycare since the birth of her little sister. She later admitted to having managed to retrieve the tablets from the kitchen cabinet, unwrap the foil, and feed the baby while her mother was asleep. To our knowledge this is the youngest case reported.

Sterilising tablets are available as paper or foil wrapped strips with dichloroisocyanurate and sodium bicarbonate as their active ingredients. They produce an exothermic and effervescent reaction in contact with water, resulting in production of hypochlorite. The combined thermal and chemical injury in situ causes severe oedema of the epiglottis and arytenoids, resulting in airway obstruction. In both our case and a previous report,³ an enthusiastic older sibling was able to unwrap the tablets and feed the baby.

There is an urgent need to deliver these tablets in childproof containers and increase parental awareness in order to prevent potentially fatal complications.

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