Bloodless treatment of infants with haemolytic disease

It was interesting to read an excellent review in the January 2004 edition of Archives devoted to the topic of blood transfusion.1 At the same time the issues of haemolytic disease in the newborn (HDN) and alternatives to exchange transfusion (ET), were treated as follows: “A recent systematic review has shown that fewer infants require exchange transfusion for haemolytic disease of the newborn when high dose intravenous immunoglobulin is used”. Neonatologists generally applauded the efforts made in an attempt to achieve a “bloodless” solution to the treatment of Rh and/or ABO HDN in a newborn whose parents are Jehovah’s Witnesses.

In 1999 we published a case of an ABO incompatible term infant girl born to parents who were Jehovah’s Witnesses. The infant was admitted to our neonatal unit with a high serum bilirubin level necessitating ET. The parents signed a request that blood should not be administered under any circumstances. However, they authorised the use of alternative treatments: orally administered D-penicillamine (DPA) (300 mg/kg per day divided into three doses over three days), phototherapy, intravenous fluids, and recombinant human erythropoietin (200 U/kg subcutaneously on every second day for two weeks). Furthermore, we reported the outcome of this infant, who was discharged from the unit in good health following treatment. Her physical growth and motor milestones at 3 years of age revealed no red flags for neurodevelopmental maturation. In addition, the follow up audiometric tests performed on this infant were normal. To our knowledge, this was the first case of an infant who received such a combined alternative (and “bloodless”) treatment for serious ABO HDN. As far as the mechanisms of action of DPA and penicillamine in neonates, the drug does not seem to have gained acceptance in the international neonatal community. The lack of “acceptance” of DPA treatment seems sadly parochial to us, because this therapy has been used extensively in Hungary for nearly 30 years. In our own experience, more than 20 000 neonates have been treated without side effects.

The successful use of erythropoietin in the treatment of severe anaemia in a neonate, reported in our paper,1 should also be of considerable practical interest to your readers.

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References

Ethnic group differences in overweight and obese children with type 1 diabetes mellitus

We read with interest the paper by Saxena et al who report differences in prevalence of overweight and obesity in children of different ethnic groups.1 Overall, 35% (n = 53) of children with type 1 diabetes in our centre were either overweight (>91st centile on BMI charts1), or obese (>95th centile), with 18% (n = 27) of the total being obese. This compared to 23% overweight and 6% obese, respectively, in the study by Saxena et al. None of the children under the age of 4 years were overweight/obese. All the other three age groups from our service showed a higher prevalence of obesity compared to the data from Saxena et al (table 1). There was no significant difference in the proportion of overweight (19% v 16%, p = 0.57) or obesity (16% v 20%, p = 0.57) between girls and boys.

There were no statistically significant differences in the rates of overweight or obesity between white Caucasian and South Asian children at any age grouping. Furthermore, there was no significant difference in the two subgroups in relation to age, duration of diagnosis, daily insulin requirement, and metabolic control (median HbA1c 8.4% v 8.8% respectively).

In conclusion, just as there is a worrying high and increasing level of overweight and obesity in the general population,1 we have confirmed that this is an even greater problem in children and adolescents with diabetes in both our major ethnic groups. The concerns expressed by Saxena and colleagues1 are even greater in children with diabetes because of the adverse cardiovascular prognosis for young people with type 1 diabetes.2

Table 1 Prevalence of obesity and overweight in children with type 1 diabetes mellitus

<table>
<thead>
<tr>
<th>Factors</th>
<th>Overweight</th>
<th>Obesity</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>n</td>
<td>m</td>
</tr>
<tr>
<td>Age group (y)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2-4</td>
<td>3</td>
<td>0</td>
</tr>
<tr>
<td>5-9</td>
<td>33</td>
<td>6</td>
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<td>10-15</td>
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<td>14</td>
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<tr>
<td>16-18</td>
<td>24</td>
<td>6</td>
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<tr>
<td>Sex</td>
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<td></td>
</tr>
<tr>
<td>Male</td>
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<td>12</td>
</tr>
<tr>
<td>Female</td>
<td>74</td>
<td>14</td>
</tr>
<tr>
<td>Total group</td>
<td>150</td>
<td>26</td>
</tr>
</tbody>
</table>
The management of childhood diabetes needs to focus not only on glycaemic control but also on efforts to prevent excessive weight gain and to reduce other cardiovascular risk factors.

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References


Given the fact that patients with type 1 diabetes have a life-long predisposition to recurrences of diabetic ketoacidosis, it is remarkable that the approach to the management of this complication is taught in a fundamentally different way in paediatrics and in adult medicine. In the former, the primary aim is to eliminate ketonaemia and ketonuria expeditiously, using a fixed dose and evidence based insulin infusion, namely, 0.1 unit/kg/h, which is maintained as long as necessary even if it entails the risk of hypoglycaemia, the latter eventually being circumvented through the infusion of intravenous glucose, given the fact that the resolution of acidocaea takes longer than the normalisation of blood glucose concentrations.

The teaching in adult medicine, conveyed through the medium of the handbook most likely to be used by junior doctors, is that normalisation of blood glucose is paramount, hence the preoccupation with a sliding scale insulin regimen targeted at the blood glucose, as opposed to a fixed dose regimen targeted at ketonaemia and ketonuria.

What this means is that, in the transition from childhood to adulthood, a diabetic will encounter a change in emphasis during the management of recurrences of ketoacidosis. I am not sure that this is right.

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References

Soy formulas and hypothyroidism

We were interested in recently published articles by Conrad and colleagues. They concluded that infants fed soy formula had a prolonged increase of thyroid stimulating hormone when compared to infants fed by non-soy formula. We have some criticisms of their study methods.

In this retrospective study there was a notable difference between the patient numbers in the soy diet group (n = 8) and non-soy diet group (n = 70). It is well known that in prospective studies in which data of two groups are compared, in order to gain statistically significant results there should be a minimum of 10 test subjects in each group and the numbers in the groups should be close. Although it is not essential to follow this rule in retrospective studies like the one of Conrad et al, the statistical reliance of the study fails since the soy diet group has eight patients whereas the other one has 70.

Secondly, in studies in which comparisons of any of body fluid parameters are made for each group, for better results, it is important that the materials must be studied in the same sessions using calibrated machines after the materials have been stored appropriately. This could not be achieved since the study was retrospective, and it is therefore inevitable that there were differences between the thyroid stimulating hormone and thyroxine results of the soy diet and non-soy diet groups. For these two reasons we think it is impossible to conclude that soy formula decreases the success of treatment in congenital hypothyroidism. We believe that further prospective controlled studies can better shed light on this topic.

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Evidence based guideline for post-seizure management

Following the publication of the guideline review “Evidence based guideline for post-seizure management in children presenting acutely to secondary care”1 we would like to clarify the following.

First, the guideline is published in its original algorithm format in a peer reviewed journal as well as being available on the PIER website (www.pier.shelf.ac.uk), complete with minor changes following the updated systematic review in 2002.

Second, the guideline was not published until it had been assessed with regard to ease of use and clinical impact. The findings of this large scale field study show its effectiveness in improvements in quality of care and are also published.1

Third, the original guideline was developed by The Paediatric Accident & Emergency Research Group, and represents many years of work. The individuals and affiliations at the time of the research are as follows:

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Ethics: the third dimension

In essence, ethics provide the guidelines for civilised human interaction. It is an evolving concept, but through the ages some accepted ethical principles crystallised. The first ethical definition focused on the individual’s responsibility towards his community, prioritising the interests of the community. However, the events preceding the French revolution and the brutality of the two world wars emphasised the need to protect individuals and minority groups against abuses of power. The ethical focus shifted from individual responsibility towards the protection of individual human rights. With the swing of the pendulum, individual rights were often protected to the detriment of the larger community.

In medicine the same shift in emphasis forced the current ethical debate on the delicate balance between the interests of the individual and that of the community, especially in resource limited settings. The reality of the third millennium is that all the world’s inhabitants are essentially part of the same global community. The two dimensional balance between the individual and the community need to reflect this global ethical responsibility.

The third millennium also confronts us with the neglected third dimension of our ethical responsibility. It is not only the interests of the individual versus that of the community that
Splenectomy in cystic fibrosis patients

A recent article,1 a commentary,2 and two letters3 4 in Archives have revealed controversy over the place of partial splenectomy in portal hypertension in cystic fibrosis (CF). We wish to contribute to the debate with a case report:

Our male patient was homozygous for the ΔF508 mutation. He was pancreatic insufficient, his lungs were colonised with Pseudomonas aeruginosa from an early age, and he had two episodes of allergic bronchopulmonary aspergillosis. When he was 8 years old, abdominal ultrasound showed variable echogenicity of the liver compatible with cirrhosis with thick bile in the biliary tree. Treatment with ursodeoxycholic acid was commenced. Recent upper abdominal pain associated with severe gastro-oesophageal reflux led to an anti-reflux procedure being performed when he was 9 years old. A gastrostomy button was placed at the same time for night time supplementary feeding. Cirrhosis of the liver was confirmed intraoperatively. Over the next few years a massive splenomegaly developed. Full blood count showed features of hypersplenism but he remained asymptomatic with respect to the haematological abnormality. At the age of 13 years he developed severe abdominal pain in the area over the spleen. Oral analgesia was not sufficient to deal with this ongoing pain and he was unable to attend school. He was referred to do chest physiotherapy over a number of months. He had two episodes of probable melanoma. He developed severe, intercurrent shoulder tip pain secondary to diarrhoeal irritation from splenic infarcts. Computed tomography examination of the abdomen showed the spleen’s span to be 30 cm, with two infarcts. Opiates were given to control pain but it proved to be intractable in an otherwise stoical patient. Eventually, because of the risk to his lungs, his poor quality of life and the risk posed to his gastroscopy by the massive spleen, partial splenectomy and possible splenorenal shunt were planned. Pneumococcal vaccine was withheld. His white cell count (WCC) was 1.5×10^9/L, platelet count 58×10^9/L, and INR 1.6. At laparotomy, perisplenic peritonitis in the dia-

phragmatic area necessitated a total splenectomy. Shunting was not undertaken. The spleen weighed 1834 g and there were numerous infarcts. Postoperatively he did well, patient controlled analgesia being used to encourage early mobilisation. Eight days later elective banding of oesophageal varices took place. Follow up endoscopy showed that this had ablated all the vessels. Two years later he no longer has abdominal pain, has not had any further infarcts, has normal liver function. The debate on the justification for removing all or part of the spleen in patients with CF and portal hypertension hinges on two considerations: indications and risks In their commentary, Kelly and de Ville de Goyet5 emphasised the risks: infection, compromising future transplantation, while questioning the indications in the cases presented by Thalhammer et al: hypersplenism and discomfort.6 In their rebuttal, Thalhammer and colleagues emphasise the hypersplenism and not the pain and discomfort described in their case reports. In their accompanying letter, Chazalette and colleagues do not mention pain as an indication. We would agree with Kelly and de Ville de Goyet that hypersplenism in the absence of significant consequences is not on its own an indication for this major procedure (we note the number of re-laparotomies required in these small series) but would emphasise that quality of life and local effects of the size of the spleen may justify the surgical and immunological risks.

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Think laterally!
I wish to emphasise the importance of thinking laterally while looking at skin marks in at-risk children in the setting of a child protection medical. It also highlights the need for us to be vigilant about simple things which can give rise to very suspicious looking skin marks. And lastly, perhaps most importantly, it highlights the extreme importance of honest, clear, unequivocal, contemporary notes, as this is what stopped this situation from becoming a risk management and complaint issue.

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Rib peristomal reaction: did you think about chest physical therapy?

Rib fractures are uncommon in infants. Child abuse must be suspected, especially when location is posterior, as explained by the lever phenomenon.7 The positive predictive value of rib fractures as an indicator of abuse is 95–100%.8 Bone fragility, severe cough, and cardiopulmonary resuscitation can cause rib fractures, and chest physical therapy (CPT) has only been mentioned in a recent retrospective series.9

From May 2000 to May 2003 we prospectively collected chest radiographs performed as a workup for bronchiolitis, and collected six cases of infants less than 2 years old for

I was asked to see a 6 year old child with learning disabilities for a child protection medical. He was under a care order, absence of signs regarding neglect. He was, however, living unsupervised with his parents.
whom lateral rib fractures or sequelae were diagnosed. With assistance from clinics, biology, radiology, and follow up, child abuse was ruled out. CPT was the only aetiology retained. It consisted in repetitive anterior cephalocaudal compressions and provoked cough, following French national consensus. Twelve of 14 fractures were located on the lateral part of the fourth to seventh ribs, none at the costovertebral junction; physiology-paths hypothesis (unpublished data) that during CPT, maximum pressure is located in the anterior mid-thorax, namely the fifth and sixth ribs, without any lever phenomenon. It is notable that 12/14 lesions consisted of periosteal reactions with no direct signs of fractures; this may relate to the hypothesis that repeated CPT leads to subperiosteal haemorrhages more than to real fractures. In conclusion, rib fractures secondary to CPT seem less unusual than initially reported. We are thus thorough in assessing a non-accidental injury.

Paediatricians must consider the devastating psychological effects of a wrong suspicion of child abuse on the entire family. The benefit of CPT in bronchiolitis should be validated. To assess diagnosis, radiologists must precisely determine the location of the fractures on the chest and along the rib, and precisely describe radiological features.

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BOOK REVIEWS

100 grey cases in paediatrics for MRCPCH
Nagi G Barakat. London: Royal Society of Medicine, 2003, £15.95 (paperback), pp 197. ISBN 1 85315 524 1

Well done. You’ve just received the letter from the college telling you of your success at passing the part one of membership. With a skip in your step you set about preparing for the part 2 written. You start to look through copious ECGs and lists of renal blood results and your registrar gives you her old picture book that you read each night while on the toilet. Things are going relatively well until you encounter your first grey case. You have heard much of this mythical creature that plagues the paediatric SHO. You try to defeat it but your efforts are pitiful in the face of such stiff opposition. Like the Sybil to your Basil it leaves you confused, frustrated, embarrassed, and downright cross—if only there was an unsuspecting guest to abuse. But fear no more, for this collection of one hundred real life grey cases is here to help.

Covering all the major topics, these cases are arranged into 10 question blocks, with each section providing a full and varied evening’s revision. The fact that the questions are real life cases, and they read as such, brings a much needed everyday relevance to the hours of study. Many of the cases are accompanied by relevant cases from the West and benefit from doing so. One aspect that I particularly like is that the style of question accurately reflects the new style of exam, with several options for each stem—most other titles have open ended “what’s the diagnosis?” questions terminating a long passage of information.

The author has a particular interest in paediatric neurology and this comes across with detailed and pertinent explanations spanning important neurological conditions. From personal experience, if you can get to grips with the neurology topics then you are halfway there. The questions concerned with infectious diseases, another area many trainees have difficulty with, is also explained well in an easy to read and absorb manner. This text provides the revising trainee with realistic and fair clinical conundrums that have much wider applications than just in an examination hall. Like any good revision text this collection of cases allows you to forget the examination hall. Like any good revision text this reflects the new style of exam, with several options for each stem—most other titles have open ended “what’s the diagnosis?” questions terminating a long passage of information.

The National Health Service in Scotland: origins and ideals, 1900–1950

At the back of an old family album is a photograph of a bewhiskered gentleman posing with his wife on the steps of a grand house with a pillared portico, surrounded by his staff including the chauffeur of the Rolls Royce that was de rigeur at that level of society. He was my great-uncle, a successful practitioner in Burslem, and his financial success contrasted with that of his brother, a Glasgow obstetrician, who lived in comfortable but by no means plutocratic style, his earning capacity restricted by time spent with indigent patients in the Duke Street and Royal Maternity Hospitals. This contrast between established private and service oriented practice encapsulates the differences between the English and Scottish medical systems in the early part of the twentieth century that are explored in some detail by the authors, and which he believes were largely responsible for the relatively easy transition from private to socialised medicine in Scotland.

Scotland of course had the added advantage of an earlier experiment in state provided medicine in the form of the Highlands and Islands Medical Service, and this is discussed in the first chapter of this book. However, the author points out that it had not a hitherto unknown advantage that is now all but forgotten. On the day I was born, 2 July 1936, the Report of the Committee on Scottish Health Services (the Cathcart Report) was published. It attracted little attention outside Scotland. The Times of that day, given to me as a birthday present, carried a half column summary of the report, with no editorial comment—contrasting with the two column signal given to “the report of the cholera commission”. This book is however much more than just a history of the English and Scottish medical services and is a perceptive work. It is not a history of the English and Scottish medical services but rather a history of the English and Scottish medical services. The author is in fact Dr Morrice McCrae, formerly of the Royal Hospital for Sick Children in Edinburgh, and well known in paediatric gastroenterology and cystic fibrosis circles. Morrice has been miraculously transformed into a distinguished historian, and his book will appeal not only to those interested in the development of state provided medicine, but to anyone interested in the social, economic, and political history of Scotland.

Finally, readers may well ask why a non-paediatric book has been sent for review to the Archives. The author is in fact Dr Morrice McCrae, formerly of the Royal Hospital for Sick Children in Edinburgh, and well known in paediatric gastroenterology and cystic fibrosis circles. Morrice has been miraculously transformed into a distinguished historian, and his book will appeal not only to those interested in the development of state provided medicine, but to anyone interested in the social, economic, and political history of Scotland.

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