Many patients who receive invasive medical treatment or diagnostic investigations undergo subcutaneous venous cannulation. In some situations a venous cannula may be in place for a few minutes or hours, as for patients undergoing general anaesthesia, sedation before surgery, noxious clinical procedures, or diagnostic radiological investigations. However, in some clinical situations long term cannulation for weeks or months may be required - in the critically ill or cancer patients receiving long term opiate infusions for pain relief or chemotherapy.

Patients and nursing staff are only too well aware of the negative aspects associated with venous cannulation, including pain and discomfort and infection. These may be underestimated and under-reported. Patients are known to complain of pain and discomfort not only during the actual procedure itself, but whilst the cannula is in place and symptoms may continue weeks and months after it has been removed. Venous cannulation is a common procedure which appears to have been the subject of few RCTs. It is an ideal subject for nursing research, as despite traditionally being a medical responsibility it is now increasingly done by nurses in their extended role.

**RCT search**

*Bandolier*, in its search for evidence on things that are common, has this month conducted searches for randomised controlled trials (RCTs) for venous cannulation. MEDLINE was searched from 1991, using the terms CATHETERISATION PERIPHERAL, RANDOM* and VENOUS and VENOUS CANNULATION and RANDOM*. This brought to light some interesting studies on this subject, and the most interesting are reviewed.

### Patient position and faint

Some patients undergoing venous cannulation faint. Can the proportion who faint be altered by simple things like sitting or lying? One study randomised 300 pre-surgical patients aged 18 to 40 years to a lying or sitting position during insertion of a venous cannula [1]. Blood pressure and heart rate were monitored and recorded before, during and for 6 minutes after the procedure.

The incidence of vasovagal symptoms (nausea, dizziness, sweaty, pallor, hot, cold, syncope) was 12.6% (95%CI 7.4 - 17.7%; 20/159) in the sitting patients and 2.1% (95%CI 0.0 - 4.5%; 3/141) in the recumbent patients. Two sitting patients (1.3%) experienced frank syncope. Symptomatic patients were more likely (39.1%) to have had a previous history of fainting than asymptomatic patients (8.3%).

If all patients underwent cannulation in the recumbent position, 87% of all vasovagal reactions could be prevented.

### Local anaesthetic for cannulation?

Pain on cannula insertion depends to some extent on needle size - but subcutaneous injection of local anaesthetic itself causes pain. Is there a point at which the pain from injection of anaesthetic is worse than the pain from venous cannulation?

Sixty patients about to undergo general anaesthesia were randomised to receive intravenous cannulation with Venflon 18, 20 and 22 gauge cannulae [2]. One hand was cannulated without anaesthetic and the other hand received a subcutaneous injection of 1% lignocaine, and this also was randomised. Patients had their eyes closed, and were asked to grade which was the more painful.

This elegant experiment clearly demonstrated that irrespective of needle size, most patients would benefit from local anaesthetic before venous cannulation.

### More painful

<table>
<thead>
<tr>
<th>Cannulation</th>
<th>Local anaesthetic</th>
</tr>
</thead>
<tbody>
<tr>
<td>18 gauge Venflon</td>
<td>19</td>
</tr>
<tr>
<td>20 gauge Venflon</td>
<td>20</td>
</tr>
<tr>
<td>22 gauge Venflon</td>
<td>17</td>
</tr>
</tbody>
</table>

In another study topical amethocaine cream was compared to EMLA cream for pain relief, when given before insertion of a venous cannula [3]. The results showed that both treatments gave good analgesia with median visual analogue pain intensity scores of about 10 mm (of 100 maximum possible).
Helpful heparin?

Does heparin, when added to the fluids infused through a peripheral catheter, reduce local catheter-related problems and extend catheter life?

A positive result for both questions came from a well conducted study of a trial which examined the effects of heparin (final concentration 1 unit/mL) or saline added to infusion fluids just before they were given to patients. This study [4] fails to say that it was randomised, though a detailed study of the methods indicates that it almost certainly was. Indistinguishable flasks of heparin and saline were given to nurses to add to intravenous fluids except phenytoin, amiodarone or aminoglycoside antibiotics where there was incompatibility with heparin.

The results showed that catheters had a longer life with heparin (by an average of 33 hours [95% CI 9 - 56 hours]), that there were fewer complications, and that when complications occurred with heparin that happened an average of 43 hours later than with saline (95% CI 6 - 80 hours).

Butterfly needle or Teflon cannula?

A 25 gauge butterfly needle has been tested against a Teflon cannula with a 26 gauge introducer needle and 24 gauge Teflon cannula in a crossover RCT [5] in palliative care for subcutaneous opiate infusions. Patients were randomised to receive either the butterfly at 45˚ or the Teflon cannula at 90˚ in the subclavicular or abdominal regions. When there were signs of toxicity - redness, swelling, tenderness, bruising or leaking - the alternate needle type was used at a different site.

Twenty patients completed both parts of the study. The mean duration of the butterfly was 5.3 days and for the Teflon cannula was 11.9 days. Patient and nursing preferences were both heavily in favour of the Teflon cannula.

Antibiotic bonding reduces intravascular catheter infection

Perhaps the most interesting RCT came from a JAMA article in 1991 [6]. Catheters were pre-treated with cationic surfactant and then an anionic antibiotic cefazolin was bonded onto the surface before insertion. The treated catheters were compared with untreated catheters in central venous sites in 93 patients and arterial catheters in 85 patients in an ITU.

Catheters were removed on the seventh day or earlier if not needed, after decontamination of the site of insertion with 70% alcohol, and then sent to the microbiology laboratory in sterile tubes. Microbial growths were sought on 2 cm sections by staff blind to the nature of the catheter.

There were 81 control catheters, 14% of which were infected. There were 97 antibiotic catheters, of which 2% were infected. Antibiotic bonding significantly reduced the cumulative risk of infection of the catheter.

The message seems to be lie down, use local anaesthetic, use heparin and Teflon cannulae for long-term use.

Dawn Carroll
Research Sister, Pain Relief Unit, Oxford

<table>
<thead>
<tr>
<th></th>
<th>Heparin</th>
<th>Saline</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total number</td>
<td>35</td>
<td>39</td>
</tr>
<tr>
<td>Catheter life (hours)</td>
<td>99</td>
<td>66</td>
</tr>
<tr>
<td>Total complications</td>
<td>12</td>
<td>24</td>
</tr>
<tr>
<td>phlebitis</td>
<td>12</td>
<td>16</td>
</tr>
<tr>
<td>extravasation</td>
<td>0</td>
<td>8</td>
</tr>
</tbody>
</table>

PROSTACYCLIN

From Dr Tim Higenbottam,
Regional Pulmonary Physiology Laboratory
Papworth Hospital

Re: DrugWatch - Prostacyclin (PGI₂)

I read with interest the article by Dr Tom Dent (Bandolier 8) who questioned the evidence that intravenous PGI₂ is an effective treatment for primary pulmonary hypertension (PPH). He encourages purchasers to enter patients into a large randomised controlled trial. The evidence of efficacy is scanty he states, and the drug at the moment can only be used experimentally. Perhaps an alternative view comes closer to reality.

How common is PPH?

Each year in England & Wales, perhaps only 40 patients are diagnosed as having PPH, not the 400 patients reported by Dr Dent. Of these only a small fraction can be identified from physiological measurements as having sufficiently poor prognosis to merit consideration for heart-lung transplantation. In this group of patients PGI₂ appears to be effective. Some seven years have been required to undertake the studies Dr Dent reported on a total of 149 patients, recruitment taking place both in the USA and the UK.

Large scale studies are therefore out of the question. This particular difficulty of studying a rare and fatal disease is emphasised by the fact that efficacies of the other current treatments for PPH have not yet been tested by randomised controlled trials. This includes the milder forms of treatment such as calcium antagonists and anticoagulants or invasive treatments such as transplant surgery.

What then can be concluded from our present state of understanding about the effects of PGI₂ in PPH patients?

Is prostacyclin an effective treatment for PPH patients?

Randomised studies over three months comparing PGI₂ with a therapy of calcium antagonists and anticoagulants showed that a physiological improvement occurred only in PGI₂-treated patients [1,2]. Of interest to clinicians caring for these greatly disabled patients quality of life also improved only in those treated with PGI₂. For those patients whose survival is limited it can be argued that PGI₂ lessens the suffering.

Does the use of PGI₂ improve survival? Dr Dent argues that differences between patients could account for the improved survival seen with PGI₂ in the British study [3]. There were, however, no significant differences in the prognostic physiological measurements between the control group and the group receiving PGI₂.

Other workers more recently have compared survival of PGI₂-treated PPH patients with the survival of PPH patients recruited in the NIH registry study [4]. They found a substantive improvement of survival in like patients, 63% in PGI₂-treated patients at 3 years compared with 41% in an historical control group (hazard ratio 2.9, CI 1.0-8.0, P=0.045 [5]). Similar historical comparisons have been used to test efficacy of transplant surgery. Why in a rare disease such as PPH should it not be equally appropriate to use this type of analytical approach to test efficacy of PGI₂?

How expensive is prostacyclin?

Dr Dent reports the cost of PGI₂ at £103 per vial. The cost to users of continuously infused drug is half this cost. Similar costs are also offered to clinicians in France to treat PPH patients. Initial costs are £25-35,000 a year, but may increase 10-fold in patients who have survived five to ten years. Such survival times would not have been predicted if the patients were not receiving the drug - perhaps a measurement of its efficacy in itself.

Who should then receive PGI₂?

By comparison with the other treatments for PPH the evidence for efficacy of PGI₂ is far from scanty, and surely can no longer be regarded as experimental. In those patients with PPH diagnosed using established criteria [4] and when physiological measurements predict a survival of less than one year, PGI₂ can lessen symptoms and improve quality of life. There is also growing evidence of the efficacy of PGI₂ in lengthening survival in these patients.

References:

**NNTs & Tinnitus**

Medical devices tend to have much less attention than drugs or other therapies. A recent article has explained beautifully the rationale behind applying the same methods to medical devices as are used for drugs to measure their effectiveness, and what needs to be done [1].

RCTs of medical devices can be found, and the calculation of numbers-needed-to-treat (NNT) can be applied to these also. NNT is a useful measure because it can be applied to any clinically relevant and useful outcome.

**Tinnitus**

Tinnitus is a prevalent condition of varying unpleasantness. About 8% of the population (nearly 4 million people) claim to experience tinnitus which causes moderate to severe annoyance and interferes with sleep. In about 0.5% of the adult population (200,000 people) tinnitus prevents them from leading a normal life.

There is a literature which indicates that electrical stimulation can reduce or abolish tinnitus. A relatively recent RCT of an external device (Therapak) which delivers a variable low frequency pulsed electromagnetic signal rich in harmonics seems to show efficacy.

The study in Liverpool examined 58 patients with severe tinnitus of at least one year’s duration and with an unselected aetiology. Patients were given either an active or a placebo device - the active device producing no discernible sensation. They used the device for one week, and had the nature of the tinnitus examined after treatment in terms of subjective response and objectively by a repeat pure tone audiogram and tinnitus pitch and loudness matching.

**Results**

<table>
<thead>
<tr>
<th></th>
<th>Improved</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Subjective</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Active</td>
<td>14</td>
<td>31</td>
</tr>
<tr>
<td>Placebo</td>
<td>2</td>
<td>23</td>
</tr>
<tr>
<td><strong>Objective</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Active</td>
<td>13</td>
<td>31</td>
</tr>
<tr>
<td>Placebo</td>
<td>5</td>
<td>23</td>
</tr>
</tbody>
</table>

**Numbers-needed-to-treat Calculation**

The calculation for NNT is:

\[
NNT = \frac{1}{(\text{improved active} / \text{total active}) - (\text{improved placebo}) / \text{total placebo})}
\]

For the **subjective** measure the NNT was:

\[
NNT = \frac{1}{(14 / 31) - (2 / 23)} = 2.7
\]

For the **objective** measure the NNT was:

\[
NNT = \frac{1}{(13 / 31) - (5 / 23)} = 4.9
\]
Conclusion

The ideal solution for people with tinnitus is for it to be abolished - and failing that for it to be improved. A simple calculation for a simple device in a short but elegant trial indicates that, compared with placebo, at least twenty people will benefit for every 100 with tinnitus who use the device. In the absence of any other successful treatment, this could be a clinically useful yield.

References:


How do I find....?

A COPY OF THAT REPORT/ARTICLE/BOOK?

You’ve done the search, or had a search done, and now you have a list of references and maybe even abstracts for some of them. You may have seen that a report has just been published on that very topic that interests you. Getting hold of the full articles or books will be relatively easy; getting hold of a report may be a little more complicated.

At first sight your local postgraduate centre library may look pretty small and unlikely to have all the journals with the articles you want. They certainly don’t have the books that you have on your list.

Don’t panic - this is not a problem

Your library will be able to supply you with almost any document you ask for from a variety of sources:-

- The library’s own stock. You (or the library staff) will be able to copy an article. Copyright has to be taken seriously, and if your librarian will not take a request over the telephone it is because a signature from you is needed to comply with the law. This will prohibit making multiple copies from an article. The librarian will, however, supply you with the necessary forms to expedite your request - by post or fax.

- Inter-library loan from other libraries in the region or the British Library (though there will usually be a fee for this service). New document delivery services mean that once you have located the reference you can request a copy of the article by post, fax or e-mail. If you are prepared to pay around $25 with your credit card, then you can order it yourself (your librarian will give you details of how to do this).

- Downloading from a full-text database (CDROM or Internet). There are a growing number of full-text reference sources on the Internet. If you are a “cybersurfer”, or want to be, read David Penchon’s excellent summary ‘Electronic Public Health’ which lists a number of health information sources and also gives some practical tips on how to get hooked up to the Net. To get hold of a copy phone David on 01223 375319 or e-mail (if you can) dcp@cix.compulink.co.uk. New health information sources are appearing every month on the Internet.

Elementary

No self-respecting library can nowadays afford to regard itself as an island! So remember that even if your library does not have a particular item, they can get it for you - and if necessary, within hours. Your library is part of a network of healthcare libraries not only in your region, but nationally and world wide.

Libraries are information nodes. They cooperate, know who has what, and can extend local services many times over. Their professional staff are the Sherlock Holmes’s of the health information world - they know how to find the evidence.

All you have to do is ask!

Judy Palmer
Health Libraries Information Network
John Radcliffe Hospital, Oxford.
Tel: 01865 221950  Fax: 01865 220040
e-mail: judith.palmer@hclu.ox.ac.uk

Stopping Smoking

Diana Sanders, of the London School of Hygiene and Tropical Medicine, published a review of smoking cessation interventions in 1992. The review was commissioned by the Health Education Authority to examine the effectiveness of interventions aimed at encouraging people to stop smoking while they are patients in the health service. Interventions in both primary care and secondary care settings were included.

The document includes an overview of the research, a critique of the evidence for effectiveness of interventions, recommendations and a detailed bibliography with abstracts of key papers.

Inclusion criteria

Published literature on smoking cessation interventions from the 1970s to 1991 were sought from clearly identified sources. Articles included in the review had to meet at least one of the following criteria:-

- a minimum of 100 subjects in each study group.
- randomisation of the intervention.
- follow up for a minimum of one year.
- validation of smoking status in some or all of the study group.
- an intervention used as part of, or in addition to routine care.
Over 100 references were identified and 68 abstracts are listed. No formal meta-analysis is included.

## Primary care

In primary care the evidence from many trials demonstrated that even brief, one-off advice from the GP to stop smoking in the context of a general consultation is effective in helping a small but statistically and clinically significant number of people to stop smoking, and remain non-smokers for at least a year. Success rates can be boosted by:

- offering smokers health education and self-help booklets
- emphasising to smokers the short-term benefits of stopping smoking rather than dwelling on long-term dangers
- negotiating a date to stop smoking
- warning smokers that they will be asked about their progress on subsequent visits
- the use of nicotine replacement therapy in selected individuals (see Bandolier 9 for a review of nicotine replacement)

Whilst GPs may be most effective in offering brief advice, practice and community nurses, or counsellors, can offer more intensive intervention and support.

## Supporting the supporters

Although brief advice and support to stop smoking may achieve long-term cessation rates of 5%, this still means that for every “success” primary care staff will offer many episodes of smoking cessation advice with little response. Thus encouragement and support of the primary care team is needed to motivate them to invest time and energy in helping smokers stop.

## General medical outpatients

In the general medical outpatient setting the small number of trials indicate that advice to stop smoking is as effective as that provided by GPs. For smokers who have diseases caused by smoking, pregnant smokers, and patients who have suffered from myocardial infarction, brief advice with follow-up in the form of letters or follow-up clinics increases the cessation rate.

This comprehensive report, which would be valuable to anyone planning a smoking cessation service, indicates that health professionals can help a significant number of individuals stop smoking, and that higher cessation rates can be achieved by increasing the intensity and level of support offered to smokers. Motivating health professionals to offer consistent help to smokers and supporting them in their efforts to encourage smokers to stop is a priority.

Dr Selena Gray
Clinical Advisor, Regional R&D Directorate, Bristol.


## GRiPPing the evidence using CRITICAL APPRAISAL SKILLS

CASP (Critical Appraisal Skills Programme) helps people develop the skills they need to appraise evidence affecting important local purchasing decisions. CASP workshops bring together a multi-disciplinary group of people such as GPs, consumers, purchasers and providers, which promotes collaboration within districts and develops a common desire to work together to get research evidence of effectiveness into practice.

## Why GRiPP the evidence?

For the last 15 years, governments world-wide have struggled to contain the costs of health services and to use their available resources more efficiently. As policy makers grappled with ways to achieve this goal the emphasis focused on cost containment and the debate about effectiveness took second place [1].

In 1993 the Minister of Health in England identified ‘a sound knowledge base’ as one of seven ‘stepping stones’ to successful purchasing. This means purchasers must be able to assess the effectiveness and efficiency of medical interventions so that resources can be used to maximise the health gain of the population. That is, they must understand the benefits, costs and adverse effects of the services which they commission.

This task is made all the more urgent by Professor Eddy’s estimate that only 15% of the medical interventions that we carry out in the NHS have been proved to be effective in improving patients’ health [2]. Even where interventions have been shown to be effective, their translation into clinical practice can take decades, for example the use of steroids in pre-term labour [3].

## What is CASP?

Purchasers frequently use reviews of evidence to give them the information they need. ‘Reviews’ are papers which synthesise the results of primary studies and so make scientific research available to a wider audience. Reviews, however, can often be poorly conducted and can be misleading in their conclusions.

To help people make sense of evidence about effectiveness,
the former Oxford RHA set up CASP. CASP is now part of the Anglia and Oxford Region’s GRiPP (Getting Research into Practice and Purchasing) project. In developing methods of helping people appraise reviews of evidence, CASP has worked closely with the UK Cochrane Centre and McMaster University in Canada.

Finding and appraising evidence about the cost effectiveness of interventions are the first steps in Getting Research into Practice and Purchasing as the CASP logo depicts.

**CASP**

CASP aims to help people develop skills in appraising evidence about clinical effectiveness through multi-disciplinary workshops. CASP is co-ordinated by a team based in Oxford. Each county in the Anglia and Oxford Region has a CASP co-ordinator who liaises and plans the workshops with the core CASP team. These individuals work only part-time for CASP.

**What are CASP workshops like?**

The workshop format is based on experience from pilot workshops and is constantly evaluated and improved. Firstly, there is an explanation about why critical appraisal skills are so important and how they fit into the GRiPP process. This is followed by an interactive talk in which the types of trials, reviews and meta-analysis, together with some basic definitions of epidemiological and statistical terms are explained. Participants then work in small groups to solve a problem scenario such as whether or not we should purchase ultra-sound screening for all pregnant women. Another scenario was whether or not dyspeptic patients who are positive for H. pylori should be treated with triple therapy. These problems are tackled by critically appraising a review article of evidence about the clinical effectiveness of that problem.

The workshops are usually multi-disciplinary which often leads to a lively debate about controversial issues. Opinions are tempered by having to critically appraise the evidence in a systematic way together.

So far 40 workshops have been held. The majority of participants (97%) have enjoyed the workshops and 90% feel that they are a good use of their time. People want more workshops in their areas and also to develop critical appraisal skills for other articles such as economic reviews and randomised control trials. They also want workshops on how to find evidence. Some participants are willing to help run future workshops and the CASP team holds training sessions for these individuals so that the workshops can be cascaded locally. We have helped people in South and West Region to run workshops and we liaise with North Thames Region where similar work is going on. We are keen to work in other areas.

**What do people get out of a CASP workshop?**

Different people get different things from a CASP workshop. This is because critical appraisal is a process which people can use in accordance with their needs. Some people only require an awareness of the importance of finding and appraising evidence, others will actually need to acquire the skills of critically appraising evidence and a few will require skills to enable them to write literature reviews.

**What are CASP’s future plans?**

Many more CASP workshops will be run in 1995. Most of these will be purchaser-based, but others are being run for consumer groups, researchers, policy makers, audit groups and board members of health authorities and NHS Trusts. It is hoped that many initiatives that plan to implement change will use CASP workshops as a springboard to successful evidence based change in practice.

If you would like more information please contact Ruairidh Milne on 01865 226741

Catherine Brogan

Ruairidh Milne

CASP
Anglia & Oxford Regional Health Authority
Old Road, Headington, Oxford.

2 Smith R. Where is the wisdom? British Medical Journal 1991 303: 798-9

**NATIONAL AUDIT OF THE NEONATAL (GUTHRIE) SCREENING PROGRAMME**

A multi disciplinary workshop was held at the Royal College of Physicians to set structure, process and outcome of a national audit of the Neonatal Screening Programme. An audit of the programme has been funded by the Department of Health.

**Neonatal screening**

In the UK the neonatal screening programme was set up in 1969 to screen blood samples of babies aged 5 -10 days old for phenylketonuria (PKU). Screening for congenital hypothyroidism was added in 1982. Locally more tests are being added to the programme e.g. screening for cystic fibrosis, amino acid disorders and haemoglobinopathies. Some of these tests may become part of the national surveillance programme. It is timely to review the screening programme twenty years after its establishment and before it is extended.
Aims

- To develop national standards for the neonatal metabolic screening programme.
- To compare practice against agreed standards nationally and to identify reasons for the failure to achieve standards.
- To identify and encourage the implementation of the changes needed to achieve the standards set.

Objectives

- To facilitate the development of widely acceptable standards against which to audit the programme.
- To determine the reported coverage of the screening programme in a range of districts and coverage by time, birth-weight, ethnic group and place of birth.
- To identify the arrangements for monitoring the screening programme at district level and to relate these arrangements to coverage findings.
- To disseminate the main findings of the audit and the recommendations for change required to achieve standards set.
- To establish agreed methods for routinely monitoring the neonatal screening programme nationally.

Proposed Standards

Outcome

- 95% of positive cases started on treatment by 21 days unless deliberately delayed for diagnostic reasons [N]
- 100% of positive cases started on treatment by 35 days unless deliberately delayed for diagnostic reasons [N]

Process

- 99% of live born infants whose births are notified to Director’s of Public Health (denominator excludes 1st week deaths)[N]
- 95% of results of first specimens available by 20 days after birth so that action may be taken [L]
- 95% of specimens received by laboratory by fourth working day after sample being taken [L]
- 90% of results available within 2 working days of receipt by lab (received by lab being day 0) [L]
- 100% of untested infants identified by 28 days of age and sample obtained within 1 week [L]

Systems

a) Routine checking against birth notification register of specimens taken on total population in place
b) Centralised and routine checking that results are received and recorded on total population in place where held
c) Written information to all parents in advance of specimen being taken
d) All results should be recorded in the parent held child record
e) Laboratories registered in routine external quality assurance schemes
f) Routine and audited recording of results at site of birth notification
g) Nominated person responsible for monitoring the programme [purchaser]
h) Nominated person to co-ordinate progress for geographical area [provider]

Mary Weston
National Screening Network, Summertown, Oxford

Facing Unwelcome Truths

Sometimes the rare and grim retains its power to shock. Bandolier was struck by the moving account of the management of children with neurodegenerative disease in a hospice for children [1]. Over 40% of the 307 children admitted to the hospice over an eleven year period had a neurodegenerative disease. A quarter of the affected families had more than one child with the disease.

The special clinical problems were communication, feeding, respiration, excess secretions, seizures, constipation, pain, movement disorders, sleep disorders and skin care. Three quarters of the children could not speak. The use of signs, initially formal then simpler, was vital for those with retained intellectual skill despite speech impairment. Again three quarters had feeding problems. Thin liquids were harder to manage than thick. The watershed decision to move to feeding by tube or by gastrostomy is clearly emotive for all involved. Respiratory distress was managed with opioids. Hyoscine patches were used to control excess secretions, and most of the 60% who had seizure problems were managed with an oral anticonvulsant regime. Constipation, affecting 40% and compounded by the liquid diet, was dealt with by a hierarchy of lactulose, senna, microenemas and then phosphate enemas.

The difficulty in identifying and treating pain in children who cannot say they hurt or describe their pain is clear. The frequent causes of the pain identified in 35% of the children were muscle spasm, joint pain and pain from gastrointestinal causes. The movement disorders (a third of the children) were managed with antiparkinsonian, muscle relaxant and anticholinergic drugs. The severe sleep disruption, children with Sanfilippo syndrome not sleeping for many consecutive nights was better managed by practical rather than drug measures. Although most of the children were immobile none developed pressure sores.

Thinking sideways the management of these rare disorders has to be based on evidence obtained in other contexts. Improving the evidence for managing movement disorders [2] and constipation is important. A second problem is how this care is provided within the context of an internal market. Reading this account the harrowing impact on the child, the family and the community comes through. What happens when this level of charitable funding is not available?

Henry McQuay
Pain Relief Unit, The Churchill, Oxford