Quality not quantity in babies

In 1975 the perinatal mortality rate for England and Wales was 19.3 per 1000. Almost certainly, the figure for 1979 will be below 15, bringing us close to the 1978 figures for France (14.7) and not very far behind Sweden (9.4).

The reason for the accelerating but delayed fall in the British perinatal mortality is not clear. It has occurred before there has been any chance to introduce measures such as monetary incentives for mothers to attend antenatal clinics or mother-and-infant surveillance clinics, the setting up of more intensive care neonatal units, and the provision of better nutrition for mothers—all campaigned for by well-intentioned reformers, including the BMJ. Nor can the improvement be due to increased monitoring of the fetus; controlled trials have shown little difference between perinatal mortality in those who were and those who were not monitored. The availability of abortion may have been a factor, since the perinatal mortality in illegitimate births is higher than legitimate births. Another factor may have been the narrowing of perinatal mortality between social classes: in 1977 the perinatal mortality in England and Wales varied from 12 per 1000 in social class I to 22 in social class V. Regions with the highest perinatal mortality have the highest proportions of both parents of social class V and of infants of low birth weight.

The perinatal mortality rate may fall a little further if there is a national amniocentesis programme to detect fetuses with lethal central nervous system abnormalities with termination of such pregnancies, but immediate prospects are poor for preventing the other important—and theoretically avoidable—cause, preterm labour. The death rate in small infants is indeed similar in Britain and the pace-setter, Sweden.

Now that the perinatal mortality in England and Wales has become similar to that in the rest of Europe, our priorities need revision. The first essential must be to ensure that wherever babies are born there is a 24-hour service for competent management of labour and resuscitation so that the small effort required in preventing asphyxia is always available. A confidential inquiry into each perinatal death would indicate where gross errors of organisation or management are occurring, but there are few paediatric pathologists in training and unless good quality postmortem studies can be arranged on a regional basis much valuable information will be lost. Of even greater importance is an inquiry about each handicapped child to determine whether there were any preventable obstetric factors. Severe cases of handicap can be detected at the age of 2 years but the milder disabilities cannot be found until the age of 5. This continuous survey could be arranged on a district basis with the district community physician and community medical specialist (child health) co-operating. But until each newborn infant has personal case notes, or at least a computer number, linking infant and obstetric care will be difficult. Research on the needs and the prognosis for infants of extremely low birth weight could be continued in selected centres. The second essential is that all mothers in preterm labour should be transferred to a unit with intensive care facilities for the newborn. A preterm infant asphyxiated in a peripheral unit cannot have his irreversible damage repaired, no matter how good the intensive care unit to which he is later transferred.

The incidence of handicap has long been assumed to run parallel with the perinatal mortality rate. This relation has been confirmed in long-term studies, and may not be applicable when the perinatal mortality rate is low. In at least one unit the handicap rate for infants weighing below 1500 g has remained at 10%, during 15 years. Infants weighing less than 750 g at a gestational age of 25 weeks can now be kept alive. Just because the techniques are available, however, these facilities should not necessarily be provided for every infant of this size. For infants weighing less than 1000 g at birth, the survival is 15-50%, in most series and the incidence of major handicap varies from about 10% to 30%. These very small babies account for about three in 1000 births, and a region might have as many as 150 such infants each year. Whether they survive or not will make little difference to the perinatal mortality but the cost (both financial and emotional) of their survival and of supporting the inevitable number who are handicapped may be thought disproportionate. Even some of the best units are reporting rates of major handicap of 20% in survivors of assisted ventilation. Would mothers accept this high risk if given a similar choice in genetic counselling?

In the late 1960s the standard practice in Britain was to subject every baby with myelomeningocele to surgery. The same experts who advocated that policy were most vehement in rejecting it when long-term studies confirmed the disadvantages, many of which could have been predicted. Are paediatricians falling into the same trap?

Cerebral atrophy or hydrocephalus?

Clinically, cerebral atrophy usually presents a picture of dementia. Focal neurological deficits are uncommon, but epilepsy may occur. Despite the absence of raised intracranial pressure, some headache may accompany the dementia, and this combination of symptoms usually prompts investigations to exclude the expanding lesion. A finding of brain atrophy, with or without compensatory ventricular enlargement, does not call for any surgical action (beyond brain biopsy in patients in whom an accurate prognosis is demanded). Any hope of improvement depends on a search, rarely successful, for a specific underlying cause such as diabetes or syphilis; in most patients the dementia is gradually, but often intermittently, progressive.

The crucial distinction that has to be made is between dementia, which is irreversible, and hydrocephalus, which is amenable to treatment. Dilatation of the ventricular system and hydrocephalus may result from obstruction to the flow of cerebrospinal fluid (CSF) either within the ventricular system or at the exit foramina from the fourth ventricle. The continuing production of CSF by the choroid plexuses leads to a rise in fluid pressure. In childhood this may be delayed (since the vault sutures are not fused) by gross expansion of the cranium, but in adults the skull is not distensible and symptoms of raised intracranial pressure develop quickly. Death is rapid if the obstruction is not removed and the pressure relieved.

The second or extracranial part of the CSF circulation includes the basal cisterns, the tentorial hiatus, and the subarachnoid space over the surface of the cerebral hemispheres, through which the CSF circulates to the arachnoid granulations and so into the superior sagittal venous sinus. Failure or obstruction of some part of this circulation is termed “external” or “communicating” hydrocephalus. A simple finding of wide cerebral sulci associated with the ventricular dilatation shown by air encephalography or by computed tomography (CT) does not, unfortunately, enable the clinician to differentiate between atrophy of the brain and obstruction to the circulation of the CSF, which might be relieved by ventriculocisternal shunting. Either early or late communicating hydrocephalus may be due to previous subarachnoid haemorrhage, head injury, or meningitis; but in most cases the cause is never proved. Furthermore, we still do not fully understand the interaction of increased intraventricular pressure and the volume and surface area of the ventricles at the start of the process and intermittently during its subsequent course. Other factors that may be relevant are the compliance of the ventricles in relation to size and their volume. Unfortunately, errors of between 20% and 30%, may occur in estimating ventricular volume by CT or radioisotope ventriculography, though the radioisotope methods are potentially more accurate.

With so many unsolved problems of pathogenesis, the investigation and management of patients who might have communicating hydrocephalus are by no means straightforward. The distinctive clinical presentation of dementia, ataxia of gait, disturbances of external ocular movements, and incontinence is said to be characteristic of communicating hydrocephalus. In such patients a finding of hydrocephalus on air encephalography associated with poor or absent filling of the cortical sulci or with gross dilatation of the larger sulci is generally accepted as evidence of either defective CSF pathways over the cerebral hemispheres or defects of absorption of CSF into the sagittal sinus. The alternative method of investigation has been to use isotope cisternography, when abnormal and prolonged accumulation of the isotope will be shown to occur within the ventricular system after injection into the lumbar CSF.

A few years ago these “dynamic” radiological methods were thought to differentiate reliably between brain atrophy and communicating hydrocephalus, and they were used (in combination with the clinical findings) to select patients for CSF shunting. The early reports of clinical improvement were encouraging, but longer follow-up has begun to cast doubt on their reliability. Furthermore, repeated estimation by CT of the ventricular size after shunting has shown no consistent correlation with clinical improvement. And as CT has become more widely available and clinicians have become reluctant to use invasive methods of investigation they are now often presented with dementing patients who may or may not have other features of communicating hydrocephalus and whose “static” CT scan shows hydrocephalus with or without enlargement of the cerebral sulci.

What can be done to improve diagnostic certainty in these circumstances? One innovation is “dynamic” sequential CT after injection of metrizamide into the lumbar CSF; but, as with other methods, its precise clinical value is unproved. The case for long-term (24-48 hours) monitoring of intracranial pressure is somewhat stronger, and at present a finding of abnormal pressure waves on a background of normal pressure is the best indicator of a good response to CSF shunting. This method of investigation, however, is invasive, and it requires apparatus and skills that are not available in all neurosurgical departments, let alone in all general hospitals.

Should, perhaps, all dementing patients whose “static” CT scan shows ventricular dilatation be offered the chance of shunting, however small the prospects of clinical improvement may be? One approach to that question is to try to compare the cost of shunting (and its complications, which include subdural haematoma) with the cost of caring for a dementing akinetic patient. The alternative is for neurologists to accept a truly critical trial of CSF shunting in all patients who fulfil certain carefully defined clinical criteria and whose static CT scans show hydrocephalus without a mass lesion. In such a trial estimates of ventricular volume and the results of isotope cisternography and “dynamic” CT would be strictly withheld from the clinicians until at least three years of neurological and psychological assessment had been completed in a