Congenital heart disease

Transporting babies with known heart disease; who, what and where?

Frances A Bu'Lock

Perspective on the paper by Browning Carmo *et al* (see page 117)

Intravenous prostaglandin therapy is well established for maintaining or re-opening the arterial duct in newborns with duct dependent congenital heart disease. An important side effect of prostaglandin therapy is its tendency to cause apnoea. Hence elective intubation and ventilation is usually considered for infants who are transferred between hospitals soon after starting such an infusion.¹ The article from Dr Browning Carmo *et al*² (see page xxx) provides an excellent justification for transporting such infants on intravenous prostaglandin without routine mechanical ventilation. However, although the authors do comment that this may also be suitable for infants with known duct dependent heart disease (ie diagnosed antenatally), they have not highlighted how many, if any, of their own study group fall within this category.

These patients are a challenging group and represent the worst end of the spectrum of congenital heart disease.³⁻⁴ Complex single ventricle lesions and severely obstructed systemic or pulmonary circulations are overrepresented compared to the spectrum of postnatally diagnosed problems. Despite this, their parents have usually already committed to active treatment by continuing the pregnancy, and hence parental investment and expectations are often high. The theoretical advantage of diagnosing duct dependent lesions antenatally is that prostaglandin therapy can be instituted from birth to maintain ductal patency and avoid the scenario of postnatal collapse associated with ductal closure. As yet it has not been possible to prove benefit from this in terms of survival figures,⁵⁻⁸ but it is clear that maintenance of ductal patency requires a much lower dose of prostaglandin than is required to reopen the duct in a collapsed infant. Thus such infants are usually started on around 3–5 ng/kg/min of Prostin in the delivery room or after prompt transfer to the neonatal unit, and the risks of apnoea should be low.

The parents of these infants have generally faced difficult information and

choices early on in the pregnancy. They are also usually well informed about the likely cardiac condition of their child by the time of birth. With prompt institution of Prostin therapy it is frequently possible to give these parents much needed 'quality time' with their long awaited newborn (however delivered) whilst transfer is arranged. Monitoring of peripheral oxygen saturation and the occasional capillary blood gas for acid-base status can be achieved without major disruption. The parents can rejoin their baby at the cardiac centre as soon as possible after transfer, having had at least some time together to fortify themselves against what is to come.

Very few paediatric cardiac centres in the UK have on-site obstetric services, and even then, most are in a separate building from the cardiac unit. Many tertiary obstetric units do not have cardiac centres in the same city. Thus if infants with antenatally diagnosed heart abnormalities are born near their home, most will require post-natal transfer. The practice of routine intubation and ventilation for transfer of these very well looking babies seems unnecessarily intrusive.

But who should transport a well, self ventilating neonate? Are scarce neonatal or cardiac transport resources better reserved for the critically ill? This certainly seems to be an issue in the UK from time to time, leading to requests for such babies to be delivered 'in house' at the cardiac centre. 'In house' delivery is generally regarded to be good practice for babies with transposition of the great arteries, as restriction of mixing at the level of the atrial septum may cause severe desaturation after birth despite adequate ductal patency.⁹ Although at present the majority of such infants are not detected antenatally and have to take their chance, those who are should be delivered where balloon septostomy is promptly available if required. A small number of infants with other severe conditions (such as arrhythmias and hydrops fetalis) may also need 'in house' delivery negotiated on an individual

basis. This can lead to conflicts for space and time at 'cardiac' obstetric units.

Imposition of 'planned' delivery at a site distant from the parental home also imposes other problems. Anxiety levels over transport times are raised even for primiparous mothers, and there is always the potential for rapid delivery 'en route'. There is an increased need for operative delivery from labour induced before term and elective caesarean delivery may be undertaken to avoid 'uncertainty'. All these carry their risks to both mother and fetus, and also impose additional costs on the delivery units associated with the cardiac centre. These seem unnecessary when it is clearly possible to transport such infants post-natally in a safe manner, as demonstrated even across the much larger distances involved in Australian transfers² compared to those in Britain.

Despite centralisation of paediatric intensive care facilities and the establishment of specialised transport teams,⁹ only one stand-alone paediatric transport team has yet received dedicated funding in the UK. Funding for neonatal transport teams has only been available for the last 2 years. A transport team for these babies requires a practitioner competent to intubate if necessary, and an understanding of the physiological consequences of mechanical ventilation of such infants is therefore still required. Such babies should be delivered within an obstetric unit with Level 2 neonatal capability in order to safely assess and stabilise them prior to transfer. The question of whether the cardiac unit or the local neonatal team should then transfer the babies remains open.

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Fear of death and dying

Fear of death and dying

Martin Ward Platt

Perspective on the paper by Barr (see page 104)

It would be possible to respond to Peter Barr's paper "Relation of neonatologists' end-of-life decisions to their personal fear of death" with anger, disbelief or indifference, and it could be misunderstood as suggesting that covert euthanasia is a rampant practice in neonatal units, at least in Australasia. Neither these responses nor such a misreading would do justice to a paper that explores the personal attitudes that mediate the decision-making processes of neonatal consultants when faced with babies in whom the possibility of forgoing continued life support has become an option to be seriously considered.

In a survey of Australian and New Zealand (ANZ) neonatologists, using validated instruments for measuring dimensions related to fear of death or dying, Barr confirmed that some ANZ neonatologists are comfortable with knowingly hastening death with sedation and analgesia. This has already been shown in Europe, and Barr's paper suggests that the finding is generalisable across culturally similar nations. The most striking finding was an association between the acknowledgement of this practice and greater "fear of the dying process" and "fear of premature death". We are left with the question as to whether this relationship would also have been true for the other 44% of ANZ neonatologists who did not respond to the survey, and whether it would have been true of other culturally similar jurisdictions.

So, how should we respond to these findings? Firstly, recognising that our own attitudes to death and dying are an important, if unspoken, dimension of

decision making at the end of life is important in its own right. Although our attitudes to death and dying are unlikely to undergo substantial change as a result of reading this paper, when we are reviewing the management of neonatal death with our colleagues, it may be helpful to acknowledge feelings as well as facts.

Secondly, it is most unlikely that these findings are limited to consultant neonatologists.

The whole team of people who work alongside families at such times will have their own attitudes and beliefs, as well as fears, that will not necessarily be the same as those of other members of the team. This of course includes families. Those of us who have been closely engaged in minute-by-minute terminal care have learnt first hand how different parents can be from one another in relation to their fears of the dying process, extreme terror in some contrasting with serenity or resignation in others. Given that one aspect of the art of terminal care lies in helping parents to manage their feelings when these threaten to destabilise the process, perhaps learning to deal with our own fears differently might also be appropriate. In a healthy and functional team, members know something of each other's capabilities, attitudes and feelings, and work with this knowledge to help the group to work optimally towards its goal. Where the goal is a "good" death, knowledge of each others' fears and feelings will help not only in achieving this primary goal but also in the secondary goal of ensuring that team members themselves are not hurt or damaged by

the death—an important consideration for junior nursing and medical staff who may have had little or no previous exposure to death in infants or children.

What of the doctrine of "double effect"? This is the notion that treatment of a terminally ill patient with powerful drugs for symptom control is justified, ethical and legal, even if administration of such treatment may shorten life, because the immediate need for treatment of symptoms is essential, a hastened death is not the intention and death at some point soon is inevitable. By contrast, hastening the onset of death for its own sake, even if the motivation for this action is the release of the patient from intolerable symptoms (eg, uncontrollable pain or respiratory failure), is euthanasia, and is at present illegal. In theory, these two approaches can be made to look quite different, but faced with a baby for whom there is general agreement that continuing intensive care is no longer appropriate, and in the knowledge that discontinuing such care is likely (although not always certain) to be a lethal act, the boundaries between these two positions can blur.

Seldom does this blurring cause any difficulty. Clinical teams and parents, faced with the facts of a baby's desperate clinical state, are usually able to converge their views to the point that the move from intensive care to palliative care is believed by all to be appropriate, and relief of symptoms, usually with sedation and analgesia, becomes the primary goal. Problems occur when the clinical team and the parents form divergent views (as has been seen in some high-profile cases of recent years¹), and also when there are powerfully conflicting views within the clinical team (as in the Leonard Arthur case²). Such divergences may sometimes arise from strong religious convictions, but they could plausibly be generated by humanist fears or beliefs in relation to death and dying.

Acknowledging these, and dealing with them openly, might be more constructive than leaving them as unspoken but disruptive influences on the planning of