LETTERS TO THE EDITOR

Scope

Heart welcomes letters commenting on papers published in the journal in the previous six months. Topics not related to papers published earlier in the journal may be introduced as a letter: letters reporting original data may be sent for peer review.

Presentation

Letters should be:

- not more than 600 words and six references in length
- typed in double spacing (fax copies and paper copy only)
- signed by all authors.

They may contain short tables or a small figure. **Please send a copy of your letter on disk.** Full instructions to authors appear in the July 1997 issue of *Heart* (page 97).

Bidirectional superior cavopulmonary anastomosis: how young is too young?

SIR,—The paper by Slavik *et al*[1] has a curious objective. It attempts "to define the lowest age at which the bidirectional superior cavopulmonary anastomosis can safely be used in infants with complex congenital heart defects". It is difficult to achieve this objective without a properly conducted clinical protocol. This does not seem to be the case in this study. So how have the authors achieved their objective? They have presumably performed bidirectional cavopulmonary anastomosis on younger and younger infants, and would have stopped at an age when the mortality would have become prohibitive.

Slavik *et al*[1] have in fact described the *experimental* use in a younger group of patients of what is a standard operation in an older age group.[2] Their patients were between three and seven weeks old and, in one, the pulmonary artery pressure was at systemic level. These would conventionally be contraindications to this operation. The authors are to be congratulated for their results, but their paper raises some important ethical issues.

If we accept that this was an experimental operation, was ethical committee permission obtained prior to embarking on this approach and was fully informed consent obtained from the parents? If so, were the parents informed that this was an experimental approach? The alternative conventional treatment in three out of four patients was the use of "a straightforward non-bypass surgical procedure with proven low morbidity and mortality".[2] Does this small series represent all the patients in this age group who have undergone this experimental approach analysed on the basis of intention-to-treat, or is this paper a description of their experience after "the learning curve"?

We fully endorse attempts at improving the patients' outcome for congenital heart disease and advancing medical knowledge as a result of properly conducted studies. These advances have almost invariably been dependent on innovation and avoidance of inflexible attitudes on the part of clinicians. Any "new" treatment will be critically compared with the current standards of treatment by everyone. Some, but not all, of the authors of this paper have, in the past, been disdainful of "complicated interventional techniques being applied to complex forms of congenital heart disease on an experimental basis". We find it illogical that some of the authors of the current paper[1] judge that new surgical experimental interventions are easily acceptable but have major difficulties in accepting transcatheter interventions.[2]

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1 Slavik Z, Lamb RK, Webber SA, Devlin AM, Keeton BR, Monro JL, Salmon AP. Bidirectional superior cavopulmonary anastomosis: how young is too young? *Heart* 1996;75:78–82.
2 Salmon AP, Keeton BR, Sethia B. Developments in interventional catheterisation and progress in surgery for congenital heart disease: achieving a balance. *Br Heart J* 1993; **69**:479–80.

This letter was shown to the authors, who reply as follows:

SIR,—We thank Qureshi *et al* for their comments. They raise important issues associated with clinical research generally and our paper on the cavopulmonary anastomosis in particular.

Between 1990 and 1995 we undertook a total of 24 consecutive cavopulmonary shunts (median age 16 months, range three weeks–64 months, including 10 infants) in this institution with one early death caused by bowel perforation in a nine week old (mortality 4·17%). This series has been reported as part of a large multicentre study.[1] The paper referred to by Qureshi *et al* concerns the four youngest patients in our series, all of whom were under two months old.

Knowing the age limits for any procedure is important and for many reasons there has been a consistent trend internationally to operate on children at a younger age (for example primary repair of tetralogy of Fallot). The cavopulmonary anastomosis has been utilised for nearly 40 years and the preoperative anatomical and haemodynamic factors associated with a good outcome (good ventricular function, normal distal pulmonary arteries, and a low pulmonary vascular resistance) are well established. There is already a large experience of the procedure in infancy and to a lesser degree in babies under six months of age.[2,3] There is evidence that postnatally pulmonary vascular resistance reaches its nadir at about three weeks of age.[4] We selected infants of three, four, six, and seven weeks of age who in whom we confirmed a low pulmonary vascular resistance, good left ventricular function, and normal distal pulmonary arteries. Mortality was zero.

Some centres may consider the application of a well established procedure to younger patients fulfilling accepted criteria to be experimental—our institution does not. The editorial referred to in their letter expressed concern relating to a high mortality in novel procedures not previously described in children of any age.

As emphasised in our discussion, despite a zero mortality in these four children, we remain cautious in making firm recommendations regarding the safe lower age limit for the cavopulmonary anastomosis.

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1 Webber SA, Howarth P, LeBlanc JG, Slavik Z, Lamb RK, Monro JL, *et al*. Influence of competitive pulmonary blood flow on the bidirectional superior cavopulmonary shunt: a multi-institutional study. *Circulation* 1995; **92**:279–86.
2 Salmon AP, Sethia B, Silove ED, Goh D, Mitchell I, Alton H, *et al*. Cavopulmonary anastomosis as long-term palliation for patients with tricuspid atresia. *Eur J Cardiothor Surg* 1989;**3**:494–8.
3 Chang AC, Hanley PL, Wernovsky G, Rosenfeld HM, Wessel DL, Jones RA, *et al*. Early bidirectional cavopulmonary shunt in young infants. Postoperative course and early results. *Circulation* 1993;**88**:149–58.
4 Rowe RD, James LS. The normal pulmonary arterial pressure during the first year of life. *J Pediatr* 1957;**51**:1–4.

Outcome of isolated congenital heart block diagnosed in utero

SIR,—I read with great interest the report by Groves *et al*[1] about perinatal outcome of isolated congenital heart block. They have been able to diagnose prenatally a very large series of patients, especially considering the accepted low incidence of the condition (1 in 15 000). They have given a wider perspective of the disease than previous neonatal series because they focused on prenatal outcome for a group of fetuses diagnosed and managed at a single institution.

They reported that heart block was related to anti-Ro or anti-La antibodies in most of their patients. They remarked that two patients in the group of 36 lacked any antibodies related to congenital heart block. Both of them had a bad outcome: one had fetal hydrops and died prenatally, and the other needed a heart transplantation after pacing was unsuccessful. Their histological studies showed interruption of the bundle of His in both cases. The authors suggest a new mutation or an incomplete family history as pathogenetic mechanism of the disease in these two patients.

We have studied 40 patients with isolated congenital heart block. Two patients were anti-Ro and anti-La negative and both had wide QRS complexes. One of them had complete atrioventricular block at birth with a ventricular rate of 45 beats/min; she had signs of congestive heart failure and was paced in the neonatal period. The other patient had advanced atrioventricular block with a heart rate of 80 beats/min at birth (first degree atrioventricular block, left anterior bundle branch block, right bundle branch block alternating with second degree or advanced heart block) which evolved to complete atrioventricular block with a wide QRS at age three years; his heart rate was noted to be 45 beats/min after he had a syncope at that age. A permanent pacemaker was implanted. Both patients remain well and thriving after one and 10 years of follow up.

The outcome of our patients has been better than the anti-Ro/SSA negative

patients reported by Groves *et al.* They show, in any case, remarkable similarities in relation to the location of the block within the atrioventricular node. They all had a wide QRS which suggests a distal block within the bundle of His. Groves *et al* performed a pathological examination of the hearts of their patients and demonstrated a distal lesion of the bundle of His of the type of nodoventricular block instead of atrioventricular block.[2] This is in accordance with the pathological findings of Ho *et al*,[3] and with the clinical data presented by Frohn-Mulder *et al*[4] who noted that the QRS width was wider in a group of anti-Ro negative patients compared with a group of anti-Ro positive children.

Pathogenic mechanism of isolated congenital heart block has been related to immune mechanisms mediated by anti-Ro or anti-La antibodies. Immune mediated damage is usually located proximal to the bundle of His. Damage of the conduction system in anti-Ro negative patients seems to be located distal to the bundle of His. This may explain a lower ventricular rate which could explain the poor outcome of Groves *et al*'s patients. Further serological and familial studies of anti-Ro negative patients may give insight into the mechanism of the disease.

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1 Groves AMM, Allan LD, Rosenthal E. Outcome of isolated congenital complete heart block diagnosed in utero. *Heart* 1996; 75:190–4.
2 Lev M, Silverman J, Fitzmaurice FM, Paul MH, Cassels DE, Miller RA. Lack of connection between the atria and the more peripheral conduction system in congenital atrioventricular block. *Am J Cardiol* 1971;27: 481–90.
3 Ho SY, Essher E, Anderson RH, Michaelsson M. Anatomy of congenital complete heart block and relation to maternal Anti Ro antibodies. *Am J Cardiol* 1986;58:291–4.
4 Frohn-Mulder IM, Meilof JF, Szatmari A, Stewart PA, Swaak TJ, Hess J. Clinical significance of maternal anti Ro/SSA antibodies in children with isolated heart block. *J Am Coll Cardiol* 1994;23:1677–81.

CORRECTION

Impact of the 1991 NHS reforms on the availability and use of coronary revascularisation in the UK (1987–1995)
Black N, Langham S, Coshall C, Parker J. Heart 1996;**76(suppl 4)**:1–31.
Data on the availability of whole-time equivalent (WTE) adult cardiac surgeons in Glasgow in 1994–95 was incorrect. There were 5·9 (not 10·9) WTE representing 3·38 (not 6·25) WTE per million population aged over 24 years (Appendix 1, page 25; fig 10, page 8). The comments on page 9 should read:
Consultant levels more than doubled in East Anglian, though the increase in South East Thames was only 27%, in Greater Glasgow only 22% and there was no increase in North Western (fig 10).
Similarly, the fifth statement on page 22 under *Objective 1* should read:
The availability of consultant surgeons showed little or no change between 1987 and 1995 in three regions but more than doubled in East Anglian.

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NOTICES

The First European Workshop on Hypertrophic Obstructive Cardiomyopathy under the auspices of the Working Groups on Myocardial Function and Cardiomyopathy of the European Society of Cardiology will take place on 31 October 1997 at the Imperial College School of Medicine, London, UK. Course fee (includes coffee, tea, lunch, and live teleconference) is £125. For further information please contact The Conference Centre (tel: 0171 351 8172; fax: 0171 376 3442; email a.c.allen@ac.ic.uk).

Practical Adult Cardiovascular Pathology Course will take place on 17 November 1997 at the National Heart and Lung Institute, London, UK. Course fee (includes coffee, tea, and lunch) is £125; £100 for juniors in training. For further information please contact National Heart and Lung Institute (tel: 0171 351 8172; fax: 0171 376 3442).