Original articles

Survival after balloon atrial septostomy for complete transposition of great arteries

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SUMMARY Mortality before surgery must be taken into account when comparing the surgical mortality of atrial redirection procedures (Mustard's or Senning's operation) and the arterial switch operation for patients with complete transposition. This is because the switch operation is usually performed within the neonatal period or early infancy but Mustard's or Senning's operation usually after 4 months of age. The outcome of balloon atrial septostomy was therefore assessed in all 102 infants with transposition of the great arteries (plus or minus associated anomalies) who underwent the procedure at our hospital in the 10 years from January 1975 to December 1984. We considered the procedure to have been unsuccessful if the patient died from any cause (including other surgical procedures) between the septostomy and subsequent interatrial repair (Mustard's operation) or arterial switch operation. Eighteen patients died, although in only two was this as a direct result of the septostomy.

Statistical analysis showed that low weight, presence of a persistent arterial duct, and coarctation of the aorta were significant risk factors. Early survival of infants with transposition of the great arteries has been dramatically improved after the introduction of balloon atrial septostomy.[1-3] Nevertheless, there is considerable attrition before definitive repair, which must be included in the prediction of overall outcome.[4-10]

After the description of Mustard's[11] and Senning's[12] operations for patients with complete transposition these surgical procedures became the standard treatment. They carry a fairly low operative risk[13] when there is an intact ventricular septum. Operative mortality and the risk of late complications are greater when there is an associated ventricular septal defect. The two operations are usually performed between the ages of 4 and 8 months, or later in patients with a ventricular septal defect who have had palliative pulmonary artery banding. The arterial switch[14 15] is now preferred in some centres, including our own. Because the arterial switch procedure is performed in the early neonatal period, when there is an intact ventricular septum, or during early infancy, when there is an associated large ventricular septal defect, mortality before definitive surgery is of paramount importance when comparing surgical results of atrial redirection or anatomic correction.

Patients and methods

Case records and catheter data of all infants with

transposition of the great arteries who underwent balloon atrial septostomy at our hospital between January 1975 and December 1984 were examined. Only infants with atrioventricular concordance and ventriculo-arterial discordance were included. Thus those with an imperforate atrioventricular valve, absent atrioventricular connection, double inlet ventricle, common atrioventricular valve or double outlet right ventricle were excluded.

Of the 114 case records of infants admitted with transposition of the great arteries (up to December 1984), eight were excluded as septostomy was not performed. In six patients an arterial switch procedure was performed. Four are alive and well. Two of these patients, who had presented late to the hospital, died in the postoperative period aged 4·5 and 16 months. One patient, with a large secundum atrial septal defect, ventricular septal defect, and coarctation, did not require septostomy, had repair of coarctation of the aorta and pulmonary artery banding at 2 months, and is alive and well after Mustard's procedure at 5 months. The remaining child with intact atrial septum, ventricular septal

defect, and left ventricular outflow tract obstruction is alive and well after systemic-pulmonary shunts at 3 months and 3·5 years. As these patients did not undergo balloon atrial septostomy they were excluded from this study, which addresses the question of outcome after septostomy. Four were excluded because the case notes could not be traced, although catheter data were available. This left 102 cases in the study.

Balloon septostomy was considered to be a success if the patient went on to a definitive procedure (usually a Mustard's operation) and a failure if they died after the procedure but before an atrial redirection procedure or arterial switch procedure whether or not an atrial septectomy (Blalock-Hanlon operation)[16] was performed. Actuarial analysis was used to investigate overall survival. Patients were withdrawn 'alive' on reaching definitive surgery.[9] Different variables were assessed retrospectively and the significance of each on survival was analysed using the Mann-Whitney U test and χ^2 test, as appropriate. Each variable was examined separately, but this does not imply that, if interaction were to change this variable and no other, survival would necessarily be altered in the manner predicted.[10]

The variables evaluated were age, weight, haemoglobin concentration at the time of septostomy, the operator, time taken for the procedure, the use of prostaglandin infusion before septostomy,

and the presence of associated anomalies. The latter included ventricular septal defect, persistent arterial duct, left ventricular outflow tract obstruction, and coarctation of the aorta. Ranges of values are shown in Table 1. The severity of left ventricular outflow tract obstruction was based on the gradient between left ventricle and pulmonary artery. It was regarded as mild if the gradient was less than 30 mm Hg and moderate to severe if higher. Gradients of up to 30 mm Hg may simply reflect excessive flow across a relatively normal left ventricular outflow tract.

Results

Of the 102 infants with transposition of the great arteries who underwent a septostomy during this period of study, 18 (18%) died before interatrial repair or arterial switch procedure (Table 2). Actuarial analysis of the data in Table 2 is illustrated in Figure 1 with confidence limits of 95%. All deaths occurred within six weeks of septostomy. Of these,

Table 2   *Time intervals from septostomy, showing number of patients withdrawn from study each week*

Interval since septostomy (weeks)	No of patients who died in this interval	No of patients withdrawn in this interval*
0–1	12	3
1–2	3	—
2–3	1	2
3–4	1	3
4–5	1	1

*Second column indicates patients withdrawn alive during this time for Mustard's operation.

Table 1   *Comparisons of variables in infants undergoing a successful or failed balloon atrial septostomy. Values are medians or No(%)*

Variables	Successful operation (n=84)†	Unsuccessful operation (n=18)†	p Value
Haemoglobin	14·50	14·80	NS*
Oxygen tension:			
Before	4·00	3·50	NS*
After	4·90	4·90	NS*
pH:			
Before	7·35	7·36	NS*
After	7·38	7·30	NS*
Duration of procedure (mins)	65·00*	67·50*	NS*
Age (days)	4·50	2·50	NS*
Weight (kg)	3·40	3·10	<0·05*
Ventricular septal defect	42 (50)	7 (39)	NS†
Patent arterial duct	42 (50)	18 (100)	<0·01‡
Coarctation	7 (8)	4 (22)	<0·025‡
Left ventricular outflow tract obstruction	4 (5)	3 (17)	NS‡

NS=Not significant.
*Mann-Whitney U test.
†Incomplete data available for haemoglobin, oxygen tension, and pH.
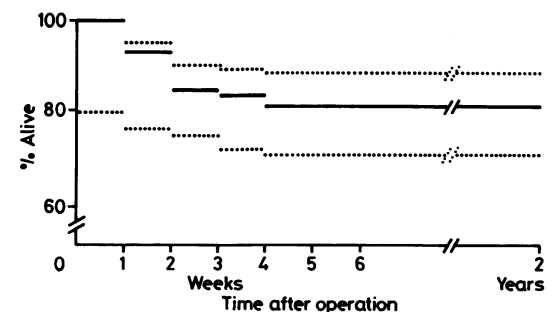‡χ^2 test.

Fig. 1   *Actuarial analysis of survival each week after balloon septostomy, with 95% confidence limits. Patients were withdrawn 'alive' at the time of a corrective procedure. Withdrawal of patients 'dead' includes only patients dying before a corrective procedure. Deaths at surgery are not included. All patients had been subjected to a further corrective operation within two years of balloon atrial septostomy.*

12 occurred within the first week and three within the second week of septostomy. There were 53 patients with complete transposition and intact ventricular septum. Ten of these (19%) died before 'corrective' surgery.

Low weight was the only measured variable that was considered to have a significant effect on outcome (p<0·05). The distribution of weight for success and failure (death) is seen in Figure 2. Histograms show a greater variance of weight in the group in whom septostomy was successful than in those in whom it was unsuccesful.

The presence of a persistent arterial duct had a deleterious effect on survival (p<0·01). Coarctation was also a significant risk factor (p<0·025). The cause of death in the 18 patients who died before interatrial repair or arterial switch is shown in Table 3. Two patients died as a direct result of the procedure and one as a result of a tear of the tricuspid valve. The other patient had a ventricular septal defect and the catheter was inadvertently passed through the ventricular septal defect and into the left pulmonary artery. This was mistaken for a pulmonary vein. Attempted septostomy resulted in a tear of the ventricular septum with acute, complete heart block and death. At necropsy, the ventricular septum had been ruptured and the atrioventricular node bisected.

In three patients balloon atrial septostomy did not result in an adequate increase in arterial oxygen tension. Elective atrial septectomy was performed, but these patients died in the perioperative period. Four patients had an apparently adequate septostomy but died after subsequent repair of coarctation of the aorta. In no way was death directly related to septostomy, but their deaths still reflect attrition

after this procedure. In our view, such patients must be included in the overall assessment of mortality.

The three patients who developed necrotising enterocolitis were all severely cyanosed and had developed a metabolic acidosis at the time of catheterisation. Whether or not enterocolitis would have developed in the absence of cardiac catheterisation and septostomy is not known. Inferior caval vein thrombosis with venous engorgement of the intestinal wall was not excluded in this group. Other causes of death included meningitis and a cerebral infarct. It is not known if these illnesses were the direct result of cardiac catheterisation, and angiography or septostomy, or both.

The patient who arrested after septostomy was already severely hypoxic and acidotic before septostomy, and death may have been the inevitable outcome whatever was attempted. This also applies to the patient who died from aspiration and the infant with progressive acidosis after a technically adequate septostomy.

It is of some encouragement to the cardiologist

Table 3 *Causes of death after balloon septostomy for the period 1974–1985. Total No of septostomies=102*

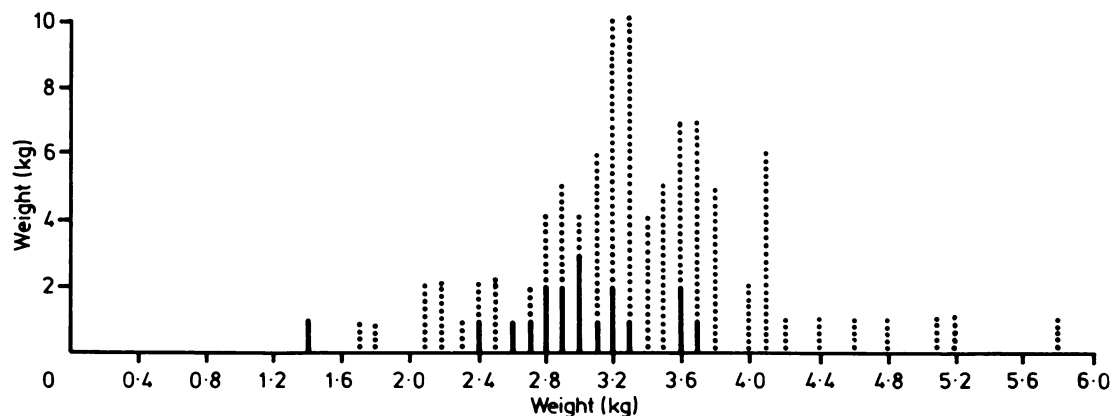|	
Post-repair coarctation ± persistent arterial duct ± pulmonary artery band	4
Necrotising enterocolitis	3
Attempted surgical septectomy	3
Cerebral infarct/meningitis	3
Cardiac arrest before septostomy	1
Aspiration	1
Severe acidosis	1
Tricuspid valve tear*	1
Ventricular septal tear*	1
Total	18

*Death directly due to septostomy.

Fig. 2 *Frequency distribution of weight at time of septostomy; success (dotted lines) and failure (continuous lines) are shown separately.*

that of the 18 deaths, only two occurred as a direct result of the procedure. The balloon atrial septostomies were performed by a total of nine different operators, and the two deaths both occurred after procedures by doctors in training, though they were supervised by a senior staff member.

Discussion

The same risk factors were evaluated in detailed analyses by Leanage et al[10] and Powell et al.[17] Our findings are in agreement with these studies and others in that coarctation and persistent arterial duct were found to be significant risk factors.[1 3 9 18 19] Analyses by Leanage and Powell also showed that age of septostomy, relative anaemia, the presence of a large ventricular septal defect, absence of left ventricular outflow tract obstruction, and pulmonary hypertension affected survival. These latter factors were found to be insignificant in our study. This may be attributed to the smaller number of patients and incomplete data for some variables. Other studies have shown that cerebrovascular accidents and cerebral abscesses occur after septostomy,[20 21] as was seen in three of our patients.

Of note was that prostaglandin E_2 infusions did not significantly affect mortality. This may indicate that the condition of the patient before the balloon septostomy does not affect the subsequent result and no patient is too sick to benefit from the procedure.[22]

The outcome of balloon atrial septostomy should be considered along with that of Mustard's or Senning's operation or the arterial switch when considering overall survival. Although it is not the purpose of this paper to comment directly on the long term results of the various operations available, we have provided sufficient information for the hypothetical considerations of the surgical approach to treatment.

We have described the actuarial survival of infants with complete transposition after balloon atrial septostomy. Most of the deaths occurred within two weeks and all within six weeks. As we have emphasised, in simple transposition of the great arteries the arterial switch procedure is performed at a younger age than Mustard's or Senning's operation. Mortality before surgery is therefore important when assessing surgical results. In patients with intact ventricular septum the neonatal switch operation is usually performed within the first two weeks of life. We do not know if this operation would have prevented all the early deaths after septostomy. It seems unlikely in that the causes of death included necrotising enterocolitis, acidosis, overwhelming infection, and cerebrovascular accidents, conditions

that may have progressed to death irrespective of any therapeutic intervention. The time in which these patients were treated largely predated widespread adoption of the arterial switch procedure for transposition both with or without a ventricular septal defect. For this reason Mustard's operation was the only corrective procedure considered in most patients.

Finally, in any hospital based series it is pertinent to ask whether the infants undergoing balloon atrial septostomy are representative of all children in the community with transposition of the great arteries.[23] It is the policy of the paediatric cardiologists at our hospital to hold joint clinics with paediatricians in district general hospitals in the hope that such contact might facilitate early recognition and transfer of patients. We believe, therefore, that most children with transposition of the great arteries from such districts are referred and the data presented here are reasonably representative.[5] It must be remembered that the results of septostomy, as with any surgical procedure, will be improved if the sickest infants die before reaching hospital. Caution is still necessary, therefore, in comparing hospital based series with population based studies, although it is encouraging to note that the outcome of balloon septostomy has improved over the last decade.[9 10]

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