Natural and modified history of complete atrioventricular septal defect—a 17 year study

P Frontera-Izquierdo, G Cabezuelo-Huerta

Abstract

We reviewed 103 cases of isolated complete atrioventricular septal defect. These cases represented 4.4% of the cases of congenital heart disease diagnosed in our hospital by catheterisation and angiography during 1971-88. Most children (n=76) had Down's syndrome. Banding of the pulmonary artery was performed in seven cases and complete repair in 67 cases. In the period 1971-82 the complete correction was performed at a mean age of 23 months with a surgical mortality of 88.8%. In the period 1983-8 the mean age at complete correction was 13 months, the mortality 43.2%, and the five year actuarial survival was 46.8%. The 22 patients that survived after complete correction were in functional classes I and II of the New York Heart Association classification. After a mean follow up of 10 years only eight (36%) of the 22 who were followed up and treated medically survived; all had developed pulmonary vascular obstructive disease and were in functional classes III or IV. Our findings stress the importance of early complete surgical repair.

Although there has been discussion on the natural history of complete atrioventricular septal defect, the general view is that all untreated cases develop obstructive pulmonary vascular disease.^{1 2} On the basis of necropsy findings Berger *et al* concluded that the survival at 6 months, 2 years, and 5 years of age is about 50, 15, and 4% respectively.³ Nevertheless, these data may not reflect the true natural history of this malformation, as many cases develop Eisenmenger's syndrome, and may survive for many years, although with a very limited quality of life.^{4 5}

Complete surgical correction offers the only present possibility of long term survival, but it is technically difficult. Since Newfeld *et al* showed that the pulmonary vascular changes may progress to fibrosis of the intima, grade III within six months, and to grade IV within a year,⁶ it appears that the surgical correction should be attempted early. In fact, in all series reported there was a high proportion of children not eligible for surgical correction because of pulmonary vascular obstructive disease.⁷⁻⁹ The vascular disease may develop faster in children with Down's syndrome, and they constitute most of the patients with this malformation.^{10 11}

We reviewed the 103 cases of isolated complete atrioventricular septal defect diagnosed and treated at the Children's Hospital 'La Fe', Valencia, since 1971 to determine more about

the development and prognosis of this malformation.

Patients and methods

The complete atrioventricular septal defect is characterised by a large atrioventricular septal defect, by a common atrioventricular valve that originates from both atria, and with deficiency of the ventricular septum.¹² Using angiohaemodynamic methods, 103 children had a complete atrioventricular defect diagnosed at this hospital from January 1971 to January 1988; these children had no other cardiac anomalies. These patients accounted for 4.4% of the 2322 children who had congenital heart defects diagnosed by cardiac catheterisation and angiography in this hospital during the period of the study. Patients with partial atrioventricular septal defects ('ostium primum') or with associated complex anomalies such as tetralogy of Fallot, left ventricular outflow obstruction, or double outlet right ventricle were excluded. Cases with associated patent ductus arteriosus and cases with the 'intermediate form' of atrioventricular septal defect, with fusion of the anterior and posterior bridging leaflets atop the ventricular septum, but with a functionally large ventricular septal defect, were included.¹² Of the 103 cases studied, 60 had the Rastelli type A, two type B, 30 type C, and 11 the 'intermediate form'.¹² Seventy six of the 103 cases (74%) were patients with Down's syndrome diagnosed cytogenetically. Seventy three of the 103 children were treated surgically (71%). The mean follow up period was 10 years.

We used the functional classification criteria of the New York Heart Association (NYHA): class I, no cardiac symptoms with ordinary activity; class II, no symptoms at rest but some limiting symptoms with ordinary activity; class III, appreciable limitation of physical activity; and class IV, cardiac symptoms at rest. The actuarial survival curves were constructed according to the modification of Anderson *et al*¹³ of the method of Kaplan and Meier.¹⁴

Results

OVERALL SURVIVAL

At the end of the follow up period, 62 patients (60%) had died, 33 (32%) survived, and eight were lost to the study.

NATURAL HISTORY

Of the 30 children treated non-surgically, in eight cases of Down's syndrome the parents

Department of
Paediatrics,
Hospital Infantil
Universitario 'La Fe',
Avda de Campanar 21,
46009-Valencia,
Spain
P Frontera-Izquierdo
G Cabezuelo-Huerta

Correspondence to:
Professor P Frontera-Izquierdo.
Accepted 14 February 1990

refused the surgical repair, the patients failed to keep subsequent appointments, and were lost to the study at a mean age of 3.5 years; their fate is unknown.

Fourteen children died (64% of the non-surgical group who were followed up): 13 within the first year (mean five months) from congestive heart failure or respiratory complications, and one at 6 years of age with Eisenmenger's syndrome. The remaining eight cases were alive at the last evaluation in 1989, had Eisenmenger's syndrome with functional class grades III or IV of the NYHA classification, and now are considered inoperable. Their clinical deterioration occurred between 1 and 3 years of life.

MODIFIED HISTORY

Of the seven children treated with banding of the pulmonary artery, three died in the immediate postoperative period, one developed obstructive pulmonary vascular disease and is not eligible for complete correction, one died during complete correction performed later, and two are awaiting complete correction.

Complete correction was performed in 67 children at a mean age of 20 months. Of these, 45 died (43 perioperatively and two late after operation). The 22 survivors of the complete repair have been followed up for a mean of 6.5 years postoperatively. The functional results are good, despite the existence of mild to moderate mitral regurgitation. In the last evaluation in 1989 their clinical state in all cases was within classes I or II of the NYHA classification.

Although the overall surgical mortality was high (65.7%; 67.2% for the complete correction) this was due in part to 88.8% mortality in the period 1971–82; in that period 36 (65%) of 55 cases diagnosed were operated on at a mean age of 23 months. Since then 37 (77%) of 48 children diagnosed have been operated on at a mean age of 13 months, with a mortality of 43.2%.

ACTUARIAL SURVIVAL CURVES

In the curve for the 103 children studied (fig 1), 23 of the children (37% of the deaths) died within the first year, only 41.9% survived to the end of the third year, and, at the end of the sixth year the survival was 28.5% and the curve approached a plateau.

Figure 2 compares the survival of the 55 children diagnosed in the period 1971–82 with that of the 48 patients diagnosed in the period 1983–8. For the former period the curve flattens after four years and only 16.4% of the patients survived. For the latter, 53.3% survived six years and 32.0% after six years. The difference in survival is significant for all points from one to five years.

The curve for the 36 children operated in the period 1971–82 (fig 3) shows that only 11.1% survived after four years. For the 37 children operated in the period 1983–8 the shape of the curve in the first years is rather similar to that for the period 1971–82, but the curve flattens off much earlier than for the former group, and

Figure 1 Combined natural and modified actuarial survival of 103 children with complete atrioventricular canal diagnosed in the period 1971–88.

Figure 2 Combined natural and modified actuarial survival of 55 children with complete atrioventricular canal diagnosed in the period 1971–82 (continuous line), and 48 children diagnosed in the period 1983–8 (dotted line).

Figure 3 Actuarial survival of 36 children with complete atrioventricular canal operated in the period 1971–82 (continuous line) and 37 children operated in the period 1983–8 (dotted line).

the survival was about 50% at two years, and 46.8% at the plateau. The differences in survival between the two groups are significant for the points between one and five years.

Discussion

The relative incidence of this cardiac malformation in our series (4.4% of the congenital heart defects diagnosed by angiohaemodynamic study) is higher than that reported in other series that include congenital heart defects diagnosed by non-invasive methods.^{15 16} This

difference probably reflects the bias of the diagnostic method used here, which favours the inclusion of patients with serious defects.

Despite the fact that 35% of the patients follow up and treated medically have survived, they are in functional class III or IV of the NYHA classification, have poor quality of life, and an unfavourable outlook, thus confirming that non-surgical management of this malformation yields very poor results.

The palliative operation adds risk to the complete correction to be performed later, does not always prevent pulmonary vascular obstructive disease (as illustrated in one of our cases), and was associated, in our study, with a high surgical mortality. Our policy at present is to use the palliative procedure only in cases with left ventricular hypoplasia.^{17 18}

Although the overall surgical mortality of the complete repair was very high, the results for the period 1983–8 were dramatically better than those for the preceding decade (1971–82). Probable causes for this improvement are the use of surgery at an early age, increase in the experience with the technique, improved perioperative care, better selection of the patients (with exclusion of those with severe pulmonary hypertension that does not decrease in the hyperoxia and tolazoline tests), and general improvement in the care of patients with Down's syndrome. Despite the high mortality of surgery in our study, the outlook is still better with surgery than without. The surgical mortality reported in other series ranged from 62% to 17%.^{12 19} Complete atrioventricular canal without major associated intracardiac anomalies can now be repaired in infants younger than 2 years in some of the leading centres with an acceptable mortality of 10–25%.^{5 7 19–21}

Prospective studies, which are required to evaluate the late results of current surgical practice, need to include more objective functional assessment. Published late non-actuarial mortality after repair of complete atrioventricular septal defect ranges from 2–19% but the duration of follow up varies considerably.^{8 12 21} There was a five year survival of about 47% in our children who were subjected to complete correction from 1983 to 1988, and good functional results of the operation. This compares with a poor prognosis both in terms of survival and of quality of life of the patients treated medically. This confirms the expectation, based on the results in other series, that complete repair drastically improves the natural history of this malformation. This also supports the opinion that this correction should be performed at an early age in as many patients as possible.^{19–22} Nevertheless, there are still doubts on the long term consequences of dysplasia of the left atrioventricular valve in the surgically corrected patient, stressing the need for monitoring of these patients over a more extended period.^{23 24}

The authors would like to thank Dr Vicente Rubio for the English version and critical reading of the manuscript.

1 Marsico F, Violini R, Calabro R, De Leva F, Caso P, Marsico L. Atrioventricular septal defects. Natural history and clinical picture. In: Quero Jimenez M, Arteaga Martinez M, eds. *Paediatric cardiology. Atrioventricular septal defects*. Madrid: Norma Ediciones, 1988:194–202.

- 2 Hoffman JIG, Rudolph AM, Heyman MA. Pulmonary vascular disease with congenital heart lesions: pathologic features and causes. *Circulation* 1981;64:873–6.
- 3 Berger TJ, Blackstone EH, Kirklin JW, Barger LM, Hazelrig JB, Turner ME. Survival and probability of cure without and with operation in complete atrioventricular canal. *Ann Thorac Surg* 1979;27:104–11.
- 4 Bull C, Rigby ML, Shinebourne EA. Should management of complete atrioventricular canal defect be influenced by coexistent Down syndrome? *Lancet* 1985;i:1147–9.
- 5 Bender HW, Hammon JW, Hubbard SG, Muirhead J, Graham TP. Repair of atrioventricular canal malformation in the first year of life. *J Thorac Cardiovasc Surg* 1982;84:515–22.
- 6 Newfield EA, Sher M, Paul MH, Nikaidoh H. Pulmonary vascular disease in complete atrioventricular canal. *Am J Cardiol* 1977;39:721–6.
- 7 Clapp SK, Burton LP, Farooki ZQ, et al. Surgical and medical results of complete atrioventricular canal: a ten year review. *Am J Cardiol* 1987;59:454–8.
- 8 Mavroudis C, Weinstein G, Turley K, Ebert PA. Surgical management of complete atrioventricular canal. *J Thorac Cardiovasc Surg* 1982;83:670–9.
- 9 Berger TJ, Kirklin JW, Blackstone EH, Pacifico AD, Kouchoukos NT. Primary repair of complete atrioventricular canal in patients less than 2 years old. *Am J Cardiol* 1978;41:906–13.
- 10 Chi TPL, Krovetz LJ. The pulmonary vascular bed in children with Down's syndrome. *J Pediatr* 1975;86:533–8.
- 11 Laursen HB. Congenital heart disease in Down's syndrome. *Br Heart J* 1976;38:32–8.
- 12 Feldt RH, Porter CJ, Edwards WD, Puga FJ, Seward JB. Defects of the atrial septum and the atrioventricular canal. In: Adams FH, Emmanouilides GC, Riemenschneider TA, eds. *Heart disease in infants, children, and adolescents*. 4th Ed. Baltimore: Williams and Wilkins, 1989:170–89.
- 13 Anderson RP, Bonchek LI, Grunkemeier GL, Lambert LE, Starr A. The analysis and presentation of surgical results by actuarial methods. *J Surg Res* 1974;16:221–30.
- 14 Kaplan EL, Meier P. Non parametric estimation from incomplete observations. *Journal of the American Statistical Association* 1958;53:457–82.
- 15 Hoffman JIE, Christianson R. Congenital heart disease in a cohort of 19502 births with long-term follow-up. *Am J Cardiol* 1978;42:641–7.
- 16 Keith JD. Prevalence, incidence and mortality. In: Keith JD, Rowe RD, Vlad P, eds. *Heart disease in infancy and childhood*. 3rd Ed. New York: Macmillan, 1978:3–13.
- 17 Silverman N, Levitsky S, Fisher E, Dubrow I, Hastreiter A, Scaglotti D. Efficacy of pulmonary artery banding in infants with complete atrioventricular canal. *Circulation* 1983;68(suppl II):148–53.
- 18 Mair DD, Mc Goon DC. Surgical correction of atrioventricular canal during the first year of life. *Am J Cardiol* 1977;40:66–9.
- 19 Chin AJ, Keane JF, Norwood WI, Castaneda AR. Repair of complete common atrioventricular canal in infancy. *J Thorac Cardiovasc Surg* 1982;84:437–43.
- 20 Folia RP, Muldre DG. Surgical correction of complete atrioventricular canal. *Am J Surg* 1987;136:121–7.
- 21 McGrath LB, Gonzalez-Lavin L. Actuarial survival, freedom from reoperation, and other events after repair of atrioventricular septal defects. *J Thorac Cardiovasc Surg* 1987;94:582–90.
- 22 Bull C, Shinebourne EA. Complete atrioventricular septal defect. Medical versus surgical treatment (1981–1983). In: Quero Jimenez M, Arteaga Martinez M, eds. *Paediatric cardiology. Atrioventricular septal defects*. Madrid: Norma Ediciones, 1988:346–52.
- 23 Studer M, Blackstone EH, Kirklin JW, et al. Determinants of early and late results of repair of atrioventricular septal (canal) defects. *J Thorac Cardiovasc Surg* 1982;84:523–42.
- 24 Rizzoli G, Mazzucco A, Brumana T, et al. Operative risk of correction of atrioventricular septal defects. *Br Heart J* 1984;52:258–65.

Commentary

In atrioventricular septal defect and other cardiac lesions sufficient relief from the symptoms of cardiac failure, with prevention of secondary changes in the heart and lungs, may not be afforded by medical treatment alone. Early surgical intervention, despite the complexity of the lesion, to induce immediate improvement and minimise long term changes in both myocardial and pulmonary vasculature is the clear message in this paper.

An additional issue raised by the authors relates to the management of children with multiple handicaps. Society today expects that each handicap will be assessed in its own right, and