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

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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 al13 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
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 complica-
tions, and one at 6 years of age with Eisen-
menger'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 clini-
cal 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 imme-
diate 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 correc-
tion) 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 chil-
dren 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% sur-
vived 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

Discussion
The relative incidence of this cardiac malforma-
tion 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. This

Figure 1 Combined natural and modified actuarial
survival of 103 children with complete atrioventricular canal

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).
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.\(^\text{17,18}\)

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%.\(^\text{12,19}\) 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%.\(^\text{5,7,19–21}\)

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.\(^\text{8,12,21}\) 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 dramatically 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.\(^\text{18–22}\) 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.\(^\text{23,24}\)

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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
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