Abstract

Objectives: Surgical repair of complete A-V canal defects (CAVCD) is a well established procedure which is currently performed in infancy. The aim of this study is to evaluate surgical results of correction in early infancy in comparison to older age.

Methods: From January 1985 to March 2001, 119 consecutive patients (age range 27 days to 83 months, mean 6.7 months) underwent repair of CAVCD in our Institution. Forms with unbalanced ventricles in association with Fallot’s tetralogy or heterotaxia were excluded from this series. Fifty-eight patients (49%) underwent correction before 3 months of age (Group A), and 61 patients (51%) after 3 months (Group B). Surgical repair was accomplished with a double patch technique in 100 patients (84%). Associated surgical lesions were treated simultaneously in 48 patients (40%).

Results: There were 11 operative deaths (<30 days) (two in Group A (3.4%) and nine in Group B (15%) (P = 0.05). The remaining patients were discharged home in good haemodynamic condition. Reoperation for postoperative left A-V incompetence occurred in five patients in Group A and in eight patients in Group B. There were eight late deaths (three in Group A and five in Group B), of which four were non-cardiac related. At a mean follow-up time of 80 months (range 2–184 months) 100 patients are asymptomatic and well, and free from oral medication. Echocardiographic examination showed absent or mild residual left A-V valve incompetence in 91 patients (49 in Group A and 42 in Group B) and moderate left A-V valve incompetence in nine patients (four in Group A and five in Group B). Kaplan–Meier survival estimates at 10 years were 90% for Group A and 75% for Group B. Kaplan–Meier freedom from reoperation at 10 years was 89% for Group A and 84% for Group B.

Conclusions: Our data demonstrate that repair of CAVCD under 3 months of age is the ideal approach to this malformation with a lower mortality rate at operation compared to older patients. Logistic analysis showed that an operative age ≥3 months is, compared to an age ≤3 months, an incremental risk factor for hospital mortality with an odds ratio of 4.8 (95% confidence limit 1–23.5) (P = 0.05). In the long term, freedom from reoperation for left A-V valve incompetence is higher when compared to children repaired at an older age.

Keywords: Complete A-V canal defects; Surgical treatment

1. Introduction

Surgical repair of complete A-V canal defects (CAVCD) is nowadays a well established procedure which is currently performed in infancy [1–5]. Still, there is no consensus as to the ideal timing for correcting these babies who develop early congestive heart failure within the first month of life, which is usually not controlled by medical therapy [3,6]. Congestive heart failure is associated with failure to thrive, and with repeated chest infections which are life threatening and can often contraindicate a primary repair. Pulmonary artery banding has been abandoned for many years, in favour of a ‘one-stage’ repair, apart from those cases with unfavourable anatomical variants [7].

We have reviewed our more recent experience with CAVCD to compare results between early correction (≤3 months) and older age.

2. Materials and methods

Between January 1985 and June 2001, 119 consecutive patients (52 males, 62 females) underwent surgical correction of CAVCD at the Department of Cardiovascular Surgery.
Surgery, University of Padua Medical School. The mean age at the time of the repair was 6.7 months (range 27 days to 83 months). Excluded from this series are forms with unbalanced ventricles, in association with Fallot’s tetralogy or heterotaxia. Trisomy 21 was the predominant feature, with an overall incidence of 75.6% (90 patients).

According to the classification of Rastelli et al. [8], 60 patients presented with type A anatomy (50%), six with type B anatomy (5%) and 34 with type C anatomy (29%). Nineteen patients (16%) had separate A-V valve orifices and restricted ventricular septal defect (VSD) (CAVCD intermediate type or transitional forms). Associated cardiac anomalies were present in 48 patients (40%) and included: patent ductus arteriosus in 48, atrial septal defect ‘secundum type’ in 32, multiple VSDs in five, persistent left superior vena cava in five, pulmonary valve stenosis in three, double orifice mitral valve in two, and tunnel subaortic stenosis in one. Primary complete repair was undertaken in 113 patients (95%); a previous pulmonary artery banding was applied in six patients (5%) (for low birth weight under 2.6 kg in five, and for an associated aortic arch reconstruction in one and coarctectomy in one). Coarctectomy alone was performed previously in another patient.

Fifty-eight patients underwent correction before the age of 3 months (49%) (Group A) while 61 patients (51%) were repaired at an older age (Group B). The Group A/Group B ratio varied from 0.16 (5/30 patients) in the period from January 1985 to December 1989 to 1.70 (53/31 patients) in the period from January 1990 to June 2001 (Fig. 1), and increased to 4.6 (28/6 patients) in the last 5 years.

All the patients were studied preoperatively with 2-D echocardiography and Doppler. Cardiac catheterization was performed in addition in 21 of the patients (18%) earlier in our experience, before 1986. Left A-V valve regurgitation (LAVVR) was studied by colour Doppler and evaluated with the flow convergence region (FCR) method [9]. Preoperative LAVVR was absent or mild in 53 patients of Group A (91.4%) and 48 patients of Group B (79%). A moderate to severe LAVVR was detected in 18 patients, five of Group A (8.6%) and 13 of Group B (21%) (Table 1).

Surgical repair was accomplished during hypothermic cardiopulmonary bypass, aortic cross-clamp and haemostatic cardioplegia (crystalloid before July 1995). Deep hypothermic circulatory arrest was selected in babies who weighed less than 4 kg in our earlier experience. Deep hypothermic cardiopulmonary bypass and low flow was the preferred strategy after March 1997.

In all the cases with an unrestricted VSD (100 patients, 84%) septal reconstruction was performed by means of a double-patch technique, utilizing a semilunar Gore-Tex (W.L. Gore and Assoc., Flagstaff, AZ) patch for the ventricular component and a separate autologous pericardial patch for the ‘ostium primum’, including also the ‘ostium secundum’ component, when present. Coronary sinus was routinely left on the right side of the patch, which drained with the systemic venous return. In the remaining 19 patients (16%), a restrictive VSD component was closed directly with pledgets suture. Complete left A-V valve ‘left’ closure was accomplished in 58 patients (49%), 32 (55%) in Group A and 26 (45%) in Group B; in the remaining 61 patients (51%), earlier in our experience, a trifoliate approach without ‘left’ closure was preferentially selected. Other reparative surgical techniques on the left A-V valve included septal hemi-leaflets resuspension in four, Kay-type annuloplasty [10] in three, commissuroplasty in two and double orifice closure in one. Associated cardiac lesions were treated simultaneously and consisted of patent ductus arteriosus ligation in 48 patients, atrial septal defect ‘secundum type’ separate closure patch in 32 patients, right A-V annuloplasty in five patients, main pulmonary artery plasty in three patients, multiple muscular VSD direct closure in three patients (in the remaining two patients the muscular VSDs were very small, without haemodynamic meaning

<table>
<thead>
<tr>
<th>No. of patients</th>
<th>Group A</th>
<th>Group B</th>
</tr>
</thead>
<tbody>
<tr>
<td>Preoperative LAVVR</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Absent to mild</td>
<td>53</td>
<td>48</td>
</tr>
<tr>
<td>Moderate–severe (P (Fisher) one-tail = 0.04)</td>
<td>5</td>
<td>13</td>
</tr>
<tr>
<td>Total</td>
<td>58</td>
<td>61</td>
</tr>
<tr>
<td>Follow-up LAVVR</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Absent to mild</td>
<td>49</td>
<td>42</td>
</tr>
<tr>
<td>Moderate–severe</td>
<td>4</td>
<td>5</td>
</tr>
<tr>
<td>Total</td>
<td>53</td>
<td>47</td>
</tr>
<tr>
<td>Reoperation rate (no. of patients)</td>
<td>8.9% (5)</td>
<td>15% (8)</td>
</tr>
<tr>
<td>Reoperation rate (no. of procedures)</td>
<td>11% (6)</td>
<td>17% (9)</td>
</tr>
</tbody>
</table>

* LAVVR, left atrioventricular valve regurgitation.
and were not found within coarsely trabeculated right ventricular septal wall), pulmonary valve commissurotomy in three patients, tunnel subaortic stenosis resection in one patient, and subpulmonary fibrous tissue resection in one patient.

Since February 1991, intraoperative 2-D echocardiography and Doppler has been routinely performed in all cases (epicardial under 5 kg of body weight and transoesophageal in bigger children).

Early mortality was defined as in-hospital death or death within 30 days of the operation. All patients were evaluated clinically and by means of 2-D colour and Doppler echocardiography, with calculation of the FCR normalized to the body surface area at the time of discharge and follow-up controls.

Follow-up was completed and updated in all cases, with a mean of 79.5 months (range 2–184 months).

2.1. Statistical analysis

Survival curves and freedom from reoperation for the two different age groups were calculated according to the Kaplan–Meier method. Categorical variables were analyzed with a contingency table with a two-tail Fischer exact test of the displayed proportion. A backward multivariate logistic analysis of the hospital mortality was done including all preoperative variables: age at operation (≤3 months, >3 months), sex, Down’s syndrome, degree of preoperative left atrio-ventricular valve regurgitation (absent–mild and moderate–severe), type of CAVCD, associated cardiac anomalies and retaining variables with a P value of <0.2.

Operative age was further investigated on a continuous scale utilizing simultaneous logarithmic transformation and quadratic exponentiation.

Table 3
Age effect modelled on continuous scale

<table>
<thead>
<tr>
<th>Hospital mortality</th>
<th>Odds ratio</th>
<th>Standard error</th>
<th>( P &gt; [z] )</th>
<th>95% confidence interval</th>
</tr>
</thead>
<tbody>
<tr>
<td>Square of age</td>
<td>0.94</td>
<td>0.04</td>
<td>0.1</td>
<td>0.87–1</td>
</tr>
<tr>
<td>Log of age</td>
<td>36.7</td>
<td>68.3</td>
<td>0.05</td>
<td>0.95–1412.1</td>
</tr>
<tr>
<td>Moderate–severe LAVVR</td>
<td>3.2</td>
<td>2.3</td>
<td>0.1</td>
<td>0.77–13.5</td>
</tr>
</tbody>
</table>

3. Results

Overall operative (<30 days) mortality was 9.2%. Two out of 58 patients died in Group A (3.4%); 9/61 patients died in Group B (15%) \( (P = 0.05) \). The mortality decreased significantly in the last 9 years reaching 3.1% (two patients died out of 64, \( P = 0.02 \)). Causes for early death were: Group A: low output syndrome (LOS) in one and pulmonary hypertensive crisis (PHC) in one; Group B: LOS in four, PHC in three, sepsis in two (Table 2).

Logistic analysis showed that an operative age >3 months was, compared to an age ≤3 months, an incremental risk factor for hospital mortality with an odds ratio of 4.8 (95% confidence limit 1–23.5) \( (P = 0.05) \). Logistic multivariate analysis (Table 3) showed that a moderate to severe preoperative left atrio-ventricular valve incompetence was a simultaneous incremental risk factor with an odds ratio of 3.2 ± 2.3 \( (P = 0.1) \). No other variables were retained in the multivariate model.

In this analysis age was modelled on a continuous scale. Results are graphically depicted in Fig. 2 according to left atrio-ventricular incompetence grade.

The effect of operative year on mortality was also obtained by forced inclusion of this variable in the logistic model and is shown in Fig. 3 according to operative age (≤3 and >3 months) and to left atrio-ventricular valve incompetence.

3.1. Follow-up

One hundred and eight patients are followed with periodical clinical and echocardiographic controls from 2 to 184 months (mean 79.5 months). Late death occurred in eight patients (7.4%). Four of them died from cardiac-related events, two in Group A and two in Group B. In the four remaining patients, death occurred from non cardiac-related events (pneumonia-immunodeficiency in two, leukaemia in one and from complications following a surgically-related neurological problem in one) (Table 2).

Echocardiographic examination showed absent or mild residual left A-V valve incompetence in 91 patients (49 in Group A and 42 in Group B) and moderate left A-V valve incompetence in nine patients (four in Group A and five in Group B).

Kaplan–Meier survival estimates at 10 years were 90% for Group A and 75% for Group B (Fig. 4).
3.2. Residual lesions

Absent-to-mild left A-V valve incompetence was found in 50 patients (89%) of Group A and in 43 patients (83%) of Group B. Moderate left A-V valve incompetence was present in 15 asymptomatic patients, six of Group A (11%) and nine of Group B (17%).

No other haemodynamically significant residual lesions were found at the last follow-up control.

3.3. Reoperation

Reoperation was performed in 13 patients (12%). The mean reoperation time from primary correction was 24.3 months, ranging from 20 days to 61 months (median 18 months). Five patients (8.9%) belonged to Group A and eight (15%) to Group B. Two patients (one in Group A and one in Group B) underwent a second reoperation 18 and 6 months after reoperation. The indication for reoperation was left A-V valve incompetence with a leak at the ‘cleft’ level in all cases, associated with a small residual VSD in four patients (one of Group A and three of Group B), and with a subaortic fibrous stenosis in one patient of Group B. In all cases the ‘cleft’ was closed thereby achieving a good valve competence in all cases. Both patients requiring a second reoperation had been treated by partial ‘cleft’ closure. In both we managed to achieve a good valve competence by closing the cleft completely. Other associated residual lesions were treated successfully simultaneously and included: a small residual peri-patch VSD closure in four patients (two in Group A and two in Group B) and a discrete subaortic fibrous resection in one patient (Group B). There were no deaths at reoperation. All the patients were discharged home in good condition. Natural A-V valves could be preserved in all patients. Kaplan–Meier freedom from reoperation at 10 years was 89% in Group A and 84% in Group B (Fig. 5).

4. Discussion

In more recent years, a better understanding of the surgical anatomy of CAVCD as well as the improvements in surgical techniques, myocardial protection and postoperative care have led to a steady reduction in operative mortality [1,6]. In most cases, congestive heart failure soon after the fall of pulmonary vascular resistance (fourth to sixth week of life) and the increase in QP/QS (ratio between pulmonary and systemic cardiac output) due to a large left-to-right shunting are poorly controlled by medical therapy and therefore surgical repair is beneficial soon after this time [3,6]. As demonstrated by Michielon et al. [6] repair at an early age is associated with a very good likelihood of preoperative competence of common A-V valves, despite the presence of a ‘cleft’ in the left A-V component. In fact, a competent common A-V valve seems to be less frequent with increasing age at operation. We think that the powerful stimulus of chronically elevated QP/QS plays an important role in the onset of annular dilatation and secondary left A-V valve incompetence [6]. In other words, early correction could eliminate, at least partially, the incidence of LAVVR in the postoperative follow-up period, which remains the most important factor for postoperative morbidity and mortality [11].

Indeed, in our present experience, the incidence of preoperative moderate–severe LAVVR was 8.6% in Group A and 21% in Group B. In the follow-up period, the incidence for reoperation for LAVVR was 11% in Group A as compared to 17% in Group B. Clearly, even though we have not found any statistical significance among the two groups, we can speculate that dealing with very fine and flimsy A-V valve tissue, as it is in young babies with CAVCD, early correction has not increased our rate for reoperation.

We all are concerned about the very delicate structure of the common A-V valve. However, septal reconstruction using the double-patch technique allows respect of valve architecture because it avoids division of the valve leaflets,
therefore limiting the secondary sequestraiton of tissue for valve division and reconstruction. When the aggressive manipulation of the delicate and flimsy common A-V valve tissue is avoided, the attention of the surgeon can be directed towards very early correction [6].

Severe cardiopulmonary instability necessitating preoperative catecholamine and respiratory support has been identified by others as a high risk factor for death [11–13]. Indeed, none of our patients who underwent repair under 3 months of age required any type of preoperative intensive care treatment.

The use of intraoperative 2-D echocardiography has been previously suggested as an extremely important manoeuvre to achieve an optimal repair [3,6,14]. In our experience with intraoperative 2-D echocardiography, which has been employed routinely in our Institution since 1988, we have found that epicardial interrogation of the heart before and after repair is still very useful as compared to transoesophageal, when currently available oesophageal probes are still too large for children with low body weight (less than 3–4 kg).

Preoperative cardiac catheterization is still employed in many centres to assess the pulmonary-to-systemic flow ratio and pulmonary vascular resistance [11,15,16]. We have found that when early repair is undertaken, there is no need for cardiac catheterization. A good preoperative (perhaps integrated by an intraoperative) 2-D echocardiography and Doppler can clearly depict the relevant surgical anatomy in detail, allowing the surgeon to plan the ideal surgical strategy for repair.

In our series, we have found that operative mortality is lower in children operated under 3 month of age. These values are probably significant to the two-tail Fisher exact test of the displayed proportion ($P = 0.06$).
In conclusion, repair of CAVCD can be safely performed before 3 months of age, with a lower mortality rate at operation compared to older patients. Logistic analysis showed that an operative age >3 months is, compared to an age ≤3 months, an incremental risk factor for hospital mortality with an odds ratio of 4.8 (95% confidence limit 1–23.5) ($P = 0.05$). In the long term, freedom from reoperation for left A-V valve incompetence is higher when compared to children repaired at an older age.

References


Appendix A. Conference discussion

Dr T. Tiaskal (Prague, Czech Republic): We have also an experience which is very similar, but I would like to know especially if you are doing perioperative echocardiographic examination after the repair, and then, what were the causes of regurgitation, whether it was anatomical causes or some dehiscence?

Dr Stellin: Well, as I showed on my slides, the reoperation was performed in eight patients mainly for left AV valve regurgitation, and the regurgitation was at the cleft site, especially in those patients who had a so-called trifoliate approach, so, in other words, in those patients in which we didn’t close the so-called cleft. And now routinely we like to close the cleft since we have to reoperate on most of the patients just because of leaking from the left AV valve due to the cleft regurgitation.

Dr J. Amato (Chicago, IL, USA): I guess my question was the same question that was asked previously. We tend to close the cleft in all of our patients and with eight reoperations and with moderate regurgitation on some of the patients, whether you want to call it a cleft or a commissure, we won’t get into that discussion, but I think all of them should be closed, and as you have come to the conclusion, you will close them from now on in.

Dr Stellin: If I can briefly answer, it is nice to know, and actually was not included in this study but was in the previous study published in Circulation in 1997, the left AV valve regurgitation is directly proportional to the age of the patient, and the postoperative left AV valve regurgitation is proportional to the preoperative left AV valve regurgitation. This is something that will then ensure an early correction rather than a late correction.

Dr Amato: Can I continue and ask you, were any of the reoperations done on the ones that you did close the cleft, because sometimes the cleft is closed either not enough or perhaps too much?

Dr Stellin: Yes. I am not sure I recall perfectly, but I probably do remember one patient in which the cleft was closed partially.

Dr T. Ebels (Groningen, The Netherlands): I saw that you have three patients with left ventricular outflow tract obstruction, and my question is, what was the anatomic basis of the obstruction and what did you do about it?

Dr Stellin: Well, the left AV valve obstruction that was present actually was treated in one patient, and that patient had so-called not the mitral valve impinging the LVO tract but was a subaortic ridge. So what we did at the time of the reoperation, because that patient was reoperated upon, together with I think a left AV incompetence, we just resect the subaortic ridge across the aortic valve.

Dr Ebels: Do you use hypothermic circulatory arrest for this repair or not?

Dr Stellin: I must say this, until three or four years ago it was a routine for us to use deep hypothermic circulatory arrest. Now we probably are better surgeons, so we do it routinely with deep hypothermia but low flow. So we don’t use circulatory arrest any longer. However, I don’t see much of a difference as far as the outcome results and the brain is concerned.

Dr Ebels: Well, hopefully in the long term.

Dr Stellin: Probably in the long term, you are right.

Dr S. Deebritz (Munich, Germany): We are all concerned about the delicate structure of those AV valves in the small babies. Can you comment on the surgical technique, one patch, two patch, or what we have increasingly applied is the Australian technique of suturing down the AV plane to the crest of the ventricular septal defect?

Dr Stellin: This is one of the points why many surgeons don’t like to do this, correct complete AV canal early in age, because the structures are a little bit flimsy. Now we have been employing since over 20 years the so-called double patch technique, and we are very careful just to respect the native architecture of the valve, try not to cut into the leaflets like the single patch technique; I have nothing against it, though. But our aim is just put a very small patch, a semilunar patch, to close the ventricular septal defect,
and try really to respect as much as possible the native architecture. So in this way we really achieve a good repair with most likely the left AV valve remaining competent.

**Dr Z. Al Halees** (Riyadh, Saudi Arabia): Was there any difference in the outcome between the Down’s and the nonDown’s in your series?

**Dr Stellin:** This is a question we were expecting. Actually we haven’t really reviewed the Down’s and nonDown’s syndrome, but we all know the nonDown’s syndrome have a more severe malformation with other associated defects. I remember for sure that two of them had a double orifice mitral valve, and in one we had to reoperate because of left AV valve regurgitation. But I have not analyzed those details in the short experience the difference between Down’s and nonDown’s.

**Dr D. Metras** (Marseille, France): Did you observe any difference in the postoperative mitral incompetence between the so-called C type or A type? Was that correlated to the anatomy type of the complete AV canal?

**Dr Stellin:** I don’t know. Actually I have a few more slides to just give a better outlook of my statistics. Nonetheless, the dominant type, so-called Rastelli type, was the A type.