Early Repair Of Complete Atrio-Ventricular Canal Malformations

*H.Elkady, MD., T.Salah ,MD., H. Hassanin,MD.,T Farouk,MD.,
and H. shawky, MD.*
Cardiothoracic surgery Department, Faculty of Medicine, Cairo University

Abstract

The ideal timing for repair of CAVC malformations remains controversial. Some authors advocate primary repair at the age of six months or earlier before the onset of irreversible pulmonary hypertension especially in the presence of Down syndrome. Others suggest two-stage repair consisting of pulmonary artery banding as a first stage followed by complete repair thereafter.

The aim of this work was to evaluate the results of early total repair of CAVCD before six months of age with the hypothesis that, with modern techniques, the current risks of CAVSD repair in children younger than 6 months and those older than 6 months are equal.

Patients & Methods*:* Twenty patients (11 females 9 males) below 6 months of age with isolated CAVC *(study group)* undergone primary total repair of CAVC defect. Another 20 patients (12 females & 8 males) with CAVC who undergone repair above the age of 6 months were selected for comparison *(control group)*. The mean age in the study group was 5.3 ± 0.49 & 12 ± 8.5 months in the control group. While the mean body weights were 4.7 ± 0.45 & 9 ± 2.5 Kg respectively. Nine patients of the study group (45%) and in 10 patients (50%) of the control group suffered from Down syndrome.

Results: There was no statistical difference in intra-operative and postoperative data between both groups, except for the duration of mechanical ventilation which was longer in the control group (48 ± 20 hours) than in the study group (36 ± 15 hours) and the incidence of recurrent postoperative pulmonary hypertensive crises which was higher in the control group (8 patients) than in the study group (4 patients).On the other hand the durations of ICU & hospital stay were higher in the study group (5 ± 1 & 10 ± 1.5 days respectively) than in the control group (4 ± 0.88 & 9 ± 1.27 days respectively.

Conclusion: We concluded that with modern techniques, the current risk factors for CAVSD repair in patients younger than 6 months and in those older than 6 months are equal.

Keywords: Complete atrioventricular canal defect, early primary repair, 6 months of age.

Introduction

The ideal timing for repair of CAVC remains controversial. Some authors advocate primary repair at the age of six months or earlier. Others suggest two-stage repair consisting of pulmonary artery banding as a first stage followed by complete repair thereafter.[1]

Improved surgical results after repair of CAVSD have been reported owing to refinements in surgical technique, improved myocardial protection, and a better understanding of the surgical anatomy. [2]
Also palliation with pulmonary artery banding is now seldom indicated and has been abandoned for a single-stage definitive surgical repair. Tight banding can cause severe pulmonary stenosis with myocardial hypertrophy, and loose banding can result in irreversible PVOD. [3]

CAVSDs are associated with high-flow systemic pressure in the pulmonary vasculature leading to fibrosis and intimal hyperplasia. This eventually leads to a reduction in the total cross-sectional area of the pulmonary vascular bed and development of pulmonary vascular obstructive disease (PVOD). [1]

The pulmonary vascular resistance along with the amount of AV valve regurgitation determine the onset of symptoms.If the peripheral vascular resistance is low, as it is normally at 6 weeks of Life, large left-to-right shunts develop through the septal defects. This in turn leads to signs and symptoms of congestive heart failure, which can also develop in the setting of severe AV valve regurgitation. About half of these patients, if left untreated, will die within the first year of life, usually from heart failure or respiratory tract infections. In those who survive, irreversible pulmonary hypertension develops and patients start developing cyanosis

from advanced pulmonary vascular disease, even with mild AV valve regurgitation. [4]

Our hypothesis is that CAVSD should be repaired before the onset of irreversible pulmonary hypertension especially in the presence of Down syndrome. This is preferably scheduled before the sixth month of life especially with improvements in anesthetic and intensive care as well as surgical techniques. [5]

Aim of work

The aim of this study is to evaluate early results of primary repair of complete AV canal malformations in the first 6 months of life.

Patients &methods

Twenty patients below 6 months of age with complete AV canal defects (**study group**) who undergone primary total repair were included. Another group of twenty patients with complete AV canal who undergone primary total repair above the age of 6 months were also selected for comparison (**control group**) in the period between November 2009 and August 2011.

No patient in this series had previously undergone palliative pulmonary artery banding, and all repairs were done electively.

Preoperative Parameters

All patiants were subjected preoperatively to complete history taking & full clinical examination. Routine preoperative

investigations were done with special emphasis on Echo cardiography (cardiac dimensions ,contractility , Cardiac valves, pulmonary artery pressure and the Rastelli type). Cardiac catheterization & angiography were performed when pulmonary artery pressure was systemic or near systemic to determine operability.

Intraoperative Parameters

Surgical technique

All patients were submitted to complete AV canal repair through complete (standard) median sternotomy. After heparinization, routine aorto-bicaval cannulation was done .

After going on bypass, and aorta crossclamped, the RA was opened along the anterior aspect of the AV groove. The right atrial wall was then suspended by 4/0 prolene stay sutures for better exposure.

From that step, three techniques were used for repair as follows:

(1)Single-patch technique: *(Figure 1)*

For detection of the borders of the cleft and superior bridging leaflet with respect to the crest of the inter-ventricular septum filling of the LV with cold saline was first done**.**
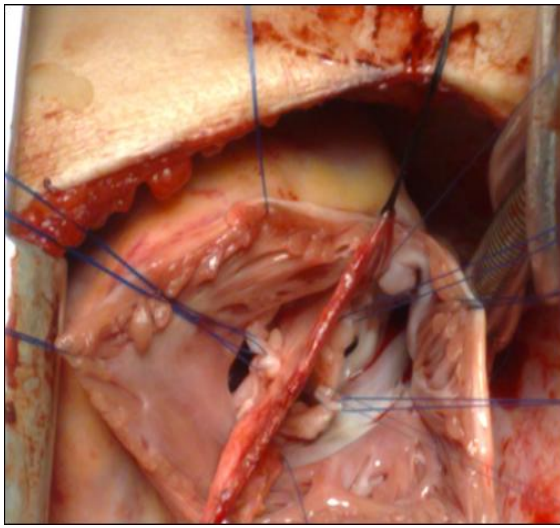
The common posterior and anterior leaflets of the AV valve were surgically divided to near the annulus (if were naturally not partitioned). The mitral portions of the common anterior and posterior leaflets were then approximated at their extremes by a marking suture. The cleft in mitral leaflets was then closed with simple interrupted sutures.

To close the VSD, mattress sutures were placed on the right ventricular surface of the upper rim of the ventricular septum. Theses sutures were passed through the lower rim of a pericardial patch, cut to conform to the size and shape of the VSD and primum ASD, and then theses sutures were tied.

Then,another mattress sutures were placed in the base of the new mitral leaflet and passed through the pericardial patch at an appropriate level to the position of the mitral valve during ventricular systole. These valve-fixing sutures were passed also through the adjacent tricuspid leaflets on the right ventricular surface of the patch. Testing of the mitral valve by saline was then done to ensure adequate repair.

While rewarming, the upper part of the pericardial patch was stitched to the atrial septum with a continuous suture to the right of the coronary sinus orifice, leaving the coronary sinus draining into the left atrium to avoid injury to the conduction tissue.

Figure (1):Single patch technique.
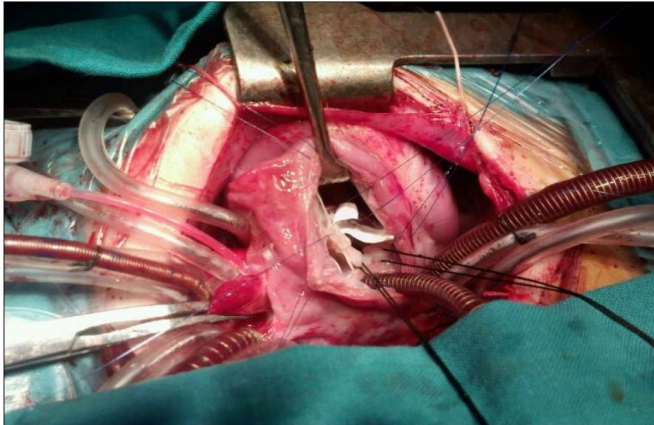
(2)Double-patch technique *(Figure2)*

Stay sutures were placed in the common anterior and common posterior valve leaflets for optimum exposure. Both valve leaflets were then inspected in order to identify the point where the common anterior and common posterior leaflets meet to form the new anterior mitral leaflet and a marking suture is placed to bring these points together. This was facilitated by injection of saline under pressure in the ventricular chambers .A crescent-shaped *Gor-tex* patch was cut to the size of the VSD whose height corresponds to the point of leaflet coaptation, and its width corresponds to the distance between the two junction-points of the interventricular septum and the AV valve annulus, at the aortic valve cephalad and AV node caudad. Running sutures were placed along the right ventricular surface of the upper margin of the ventricular septal crest and passed through the patch. Posteriorly, these sutures were placed more superficially and somewhat remotely from the rim of the VSD in order to avoid the bundle of His.

A running horizontal suture line was performed at the upper rim of the VSD patch to sandwich the AV valve leaflets over the VSD. These stitches were passed through the common anterior leaflet, the VSD patch and then the common posterior valve leaflets, respectively with the aid of the previously placed marking sutures.

At that moment interrupted sutures were used to close the cleft in the new anterior mitral leaflet and testing of the mitral valve by saline was then done to ensure adequate repair.

A second patch of pericardium was used to close the primum ASD with continuous running sutures passing through the upper rim of the VSD patch then the atrial septum, leaving the coronary sinus draining into the right atrium.

Figure (2) DOUBLE PATCH TECHNIQUT A crescent-shaped Gor-tex patch was used to close the VSD

## (3)Modified single-patch technique *(Figure 3)*

Testing, as before, was done for the detection of the borders of the cleft and the delineation of the point of coaptation of the left superior and inferior bridging leaflets.

Felted mattress sutures were placed across the top of the ventricular septum on the right ventricular surface for the VSD repair. These VSD repair stitches were passed through the AV valve leaflets in order to separate them into tricuspid and mitral components and then through the lower rim of a pericardial patch cut to the size of the ASD and finally were tied.

Multiple simple sutures were then used to close the cleft in the new anterior mitral leaflet. Then testing of the mitral valve was done by injecting saline into the left ventricle.

To complete the repair, the upper rim of a pericardial patch was attached to the atrial septum with a continuous suture leaving the coronary sinus draining into the left or right atrium.

Figure (3) Felted mattress sutures were placed across the top of the ventricular septum on the right ventricular surface for the VSD closure.
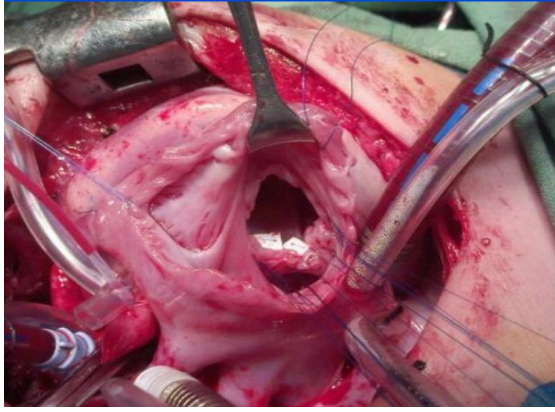
In all techniques after finishing the AV canal repair, closure of right atriotomy was performed by running 6/0 prolene sutures . Weaning from the CPB, decannulation, hemostasis and routine closure of chest were done.

Intra-Operative Parameters:

a) Cross-clamp time , total bypass time and total operative time

b) Technique used for repair: whether single-, double-, or modified single-patch technique,

c) Use of inotropes after weaning from cardiopulmonary bypass.

d) Patient rhythm on going off bypass and on discharge from the operating theatre.

Postoperative Parameters:

1- Postoperative Mechanical Ventilation time.

2- Chest tube drainage & re-exploration for bleeding:

3- Total intensive care unit stay.

4-12-lead electrocardiography was done on admission to the ICU to record the basic rhythm of the patient, and on discharge from the ICU.

5- In-hospital mortality was defined as death within 30 days of the operation

All analyses were done using the statistical software SPSS (SPSS Inc, Chicago, IL).In addition, univariate statistics using either an α2 analysis or a Fisher exact test were obtained comparing the variables: age, reoperation, mortality, Down syndrome, gender, and the degree of postoperative mitral regurgitation. A significant difference was indicated at $p < 0.05$.

Data were statistically described in terms of frequencies (number of cases), relative frequencies (percentages), mean and standard deviation values (SD). All statistical calculations were done using Microsoft excel 7 computer program (Microsoft cooperation, NY, USA).

Results

Preoperative results

In the study group (who had repair before or on 6 months), the **age** ranged from 4.5 to 6 months with a mean of 5.3±0.49 months and the **body weight** ranged from 4 to 5.5 Kg with a mean of 4.7 ± 0.45Kg.

While in the control group (who had repair after 6 months), the **age** ranged from 8 to 36 with a mean of 12± 8.5months and the **body weight** ranged from 7 to 15 Kg with of mean of 9± 2.5 Kg.

The rest of the demographic data of the patiants ,as well as the preoperative echocardiographic findings are shown in Table (1).

Table (1) Demographic data &preoperative echocardiographic data of the Patients.

Preoperative data	Study group	Control group	P Value
Demographic data **Age(Mean)**	5.3±0.49m	12± 8.5m	
Weight(Mean)	4 to 5.5 Kg	9± 2.5 Kg	
Sex **Male** **Female**	 9 (45%) 11 (55%)	 8 (40%) 12 (60%)	Non significant
Down syndrome	9 (45%)	10 (50%)	Non significant
Preoperative Echo **Rastelli type** **A** **B** **C**	 11(5%) 2 (10%) 7 (35%)	 13 (6%) 1 (5%) 6 (30%)	Non significant
VSD size(Mean)	6 ± 1mm	5±1mm	Non significant
Common AV valve regurge **Trivial-mild** **Mod.-severe**	 14 (70%) 6 (30%)	 13 (65%) 7 (35%)	Non significant
Mean PAP	60± 9 mmHg	65± 9 mmHg	Non significant

There was no statistical diferance between both groups as regards the preoperative echocardiographic findings between both groups .(P value > .05).

Intraoperative results

The intra-operative surgical data were collected in both groups and represented as Table (2)

Intra-operative data	Study group	Control group	P value
Time parameters			
Total operative time	182.5$\pm$ 8min.	190$\pm$ 8.5min.	Non significant
CPB time	122 $\pm$12min.	125 $\pm$10min.	
Cross-clamp time	88$\pm$ 7.5min.	82$\pm$ 8min	
Techniques of repair			
Double-patch	8(40%)	18 (90%)	
Single-patch	4 (20%)	2 (10%)	
Modified single-patch	8 (40%)	-	
2nd degree HB	2 (10%)	1 (5%)	Non significant

<mark>After weaning from the cardiopulmonary bypass, the patients' rhythms were recorded in all patients: In cases of the study group, sinus rhythm was restored in 18 (90%) patients, while 2 patients (10%) showed 2nd degree heart block . In cases of the control group, sinus rhythm was restored in 19 patients (95%), while 1 patients (5%) developed 2nd degree heart block. There was no statistical difference between the two groups. In all patients who developed 2nd degree heart block, pacing using temporary pace makers was done at this stage and the pace makers were transferred with the patients to the ICU.</mark>

there was no statistical difference between the two groups as regards intraoperative parameters.(P value > .05).

__Postoperative results__

All patients required postoperative mechanical ventilation, no patient was extubated in the operating theatre. The mechanical ventilation (Extubation) time in the study group ranged from 20-72 hours with a mean of 36 ± 15 hours. While in the control group, the ventilation time ranged from 24-96 hours with a mean of 48 ± 20 hours. There was statistically significant difference between the two groups (P value =.03).

Recurrent attacks of pulmonary hypertensive crises developed in 4 patients (20%) of cases of the study group, while these attacks occurred in 8 patients (40%) of cases of the control group. This was of statistically significant difference between the two groups (P value =.01). These attacks were managed by hyperventilation with higher positive airways pressures, sedatives using IV shots of Fentanyl at a dose of 3 - 5 µcg/kg or Dormicum (Midazolam) at a dose of 0.2 mg/kg and sometimes muscle relaxants using IV boluses **of Pancuronium** at a dose of 0.1mg/kg.

The total intensive care unit (ICU) stay in the study group ranged from 4-8 days with a mean of 6 ± 1 days, while the total hospital stay ranged from 15-25 days with a mean of 20 ± 2.8 days. In the control group, the total intensive care unit (ICU) stay ranged from 3-6 days with a mean of 4 ± 0.88 days, while the total hospital stay ranged from14-22 days with a mean of 18.5 ± 2.31 days which were statistically significantly lower than in patients

repaired before 6 months (P value = .02and .015 respectively). These data
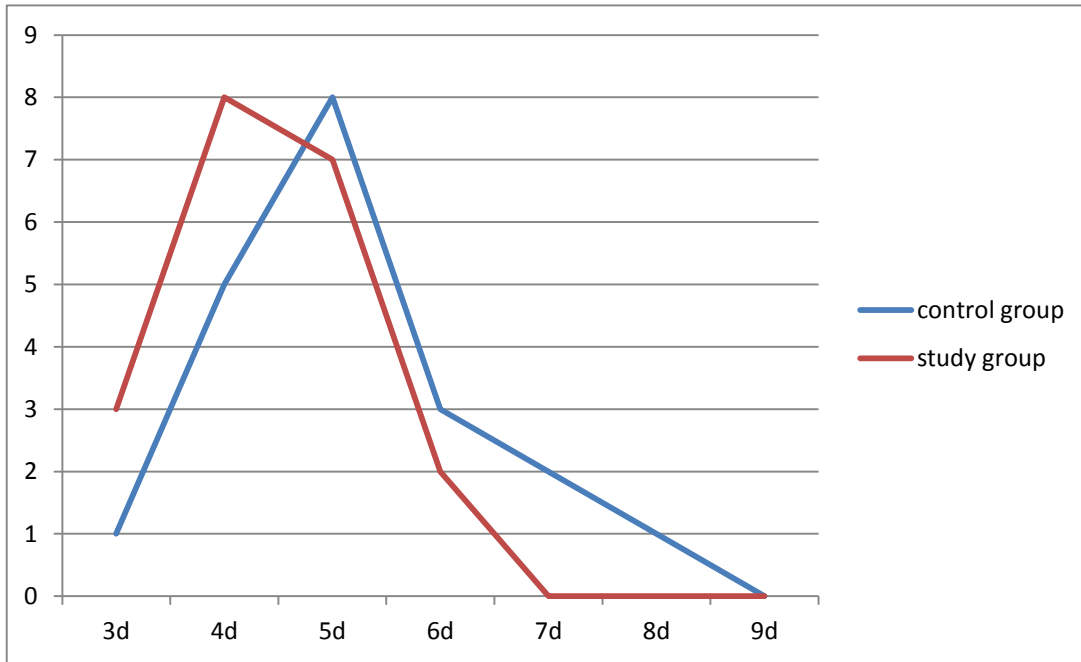are shown in curves. Figure *(4&5)*.
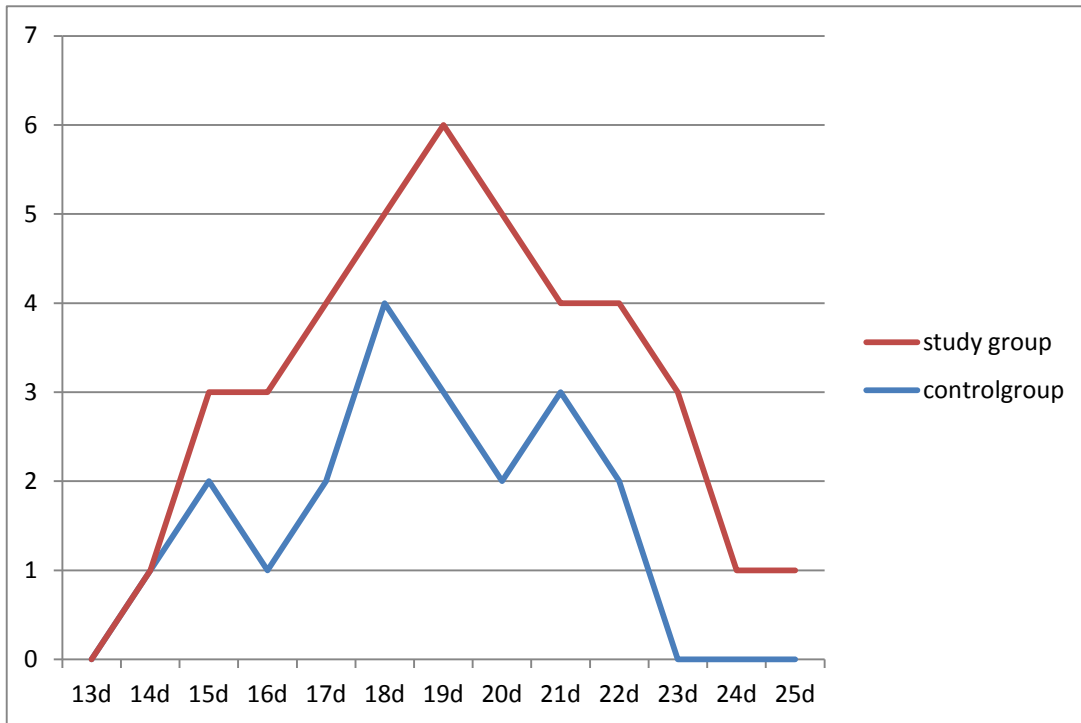

Figure (4). ICU stay duration in both groups.

Figure (5) Total hospital stay in both groups.

No hospital mortality was reported among the 20 patients of the study group. While in the control group, one patient died after a severe pulmonary hypertensive crisis during which all measures failed to restore adequate O2 saturation to keep the patient alive.

Summary of postoperative results are shown in Table (3).

Table (3). Summary of postoperative results.

Postoperative data	Study group	Control group	P value
Mechanical ventilation	36 ± 15 hours	48 ± 20 hours	Significant (.03)
Pulm. hypertensive crises	4 (20%)	8 (40%)	Significant (.01)
Chest tube drainage	60 ± 29cc	80 ± 35 cc	
Re-exploration	-	1	
ICU stay	5 ± 1 days	4 ± 0.88 days	Significant(.02)
Hospital stay	10 ± 1.5 days	9 ± 1.27days	Significant(.015)
In-hospital mortality	-	1	

Discussion

Surgical repair is the standard of care that is now offered for all patients with of CAVSD and can be performed in infancy. However, in a subset of patients, early congestive heart failure develops within the first few weeks of life that might be not controlled with medical therapy alone. It is in

these patients that controversy exists about the ideal timing and strategy of surgery. [2]

The two stage repair although was carried out to reduce in-hospital mortality and technical difficulty from operating on smaller hearts with delicate tissues, yet this approach turned out to bear all long-term complications of pulmonary artery banding .

In addition, repair of AV canal defects beyond 6 months of age has been recently shown to be an incremental risk factor for death. The chronic volume overload from left-to-right shunting increases common AV valve annular dilatation resulting in increased left AV valve regurgitation due to the difficulty in achieving good co-aptation after repair and promotes secondary pathological changes in the AV valve tissue including the cleft area as well. It is not uncommon to hear surgeons make the comment that there is 'inadequate AV valve tissue to perform valve repair'. [1]

Another sequel of delaying surgery is the development of pulmonary hypertension with elevated pulmonary vascular resistance, predisposing to postoperative pulmonary hypertensive crises. Irreversible PVOD, Heath–Edwards grade III and higher pathological changes in the lungs can occur within the first year of life, especially in the presence of Down syndrome. [6]

In Singh et al, 2006[7], study, the mean age was 2.14 months in group A who underwent repair on or before 3 months of age (26 patients) and 16.76 months in group B who underwent repair after 3 months of age (39 patients). In another study done by Stellin and colleagues, 2003, 119 consecutive patients underwent repair of CAVCD from January 1985 to March 2001. 58 patients (49%) underwent correction before 3 months of age (Group A), and 61 patients (51%) after 3 months (Group B). [8]

The lower mean age in Group A in the studies of Singh et al [7] as well as Stellin et al[8] than the mean age in our study group reflects the earlier tendency to repair complete AV canal defects, especially in the presence of intractable heart failure. While the higher mean age in group B than our control group may be attributed to the presence of more cases in group B in those studies with restrictive left to right shunts (in whom definitive repairs could be postponed).

In our study the age of 6 months was considered to be the division point between the two groups since pulmonary hypertension usually occurs by this age, while Singh and colleagues[7] as well as Stellin and colleagues[8] believed that pulmonary hypertension can occur as early as age of 3 months especially in patients with Down syndrome.

The mechanical ventilation time was longer in patients undergone repair after 6months and the incidence of pulmonary hypertensive crises was more in these patients (in whom the mean of pre-operative systolic pulmonary pressures was higher than that of patients repaired before 6months).The trend towards earlier intervention has decreased the incidence of pulmonary hypertensive crises. Although earlier reports consistently described postoperative pulmonary hypertension as a major risk factor of death, yet recent long-term results as well as our study demonstrate that early surgical intervention before the development of pulmonary vascular obstructive disease is the optimal approach. [9] In the study done by Kobayashi and colleagues, the median duration of mechanical ventilation was 11.6 hours which is much less than that in our study .This is due to the lack of Nitric Oxide gas and Extra-corporial Membrane Oxygentation (ECMO) in our intensive care units. Therefore patients are kept sedated and mechanically

ventilated for longer time for fear of the occurrence of fatal pulmonary hypertensive crises [9]

Most surgeons currently prefer to perform definitive repair before age 6 months. Reddy and colleagues propose that earlier definitive repairs do not result in an increase in the incidence of AV valve incompetence and recommend elective repair at age 2 to 3 months. In fact, early correction could partially eliminate the incidence of left AV valve regurgitation in the postoperative period as well as reoperation in older patients. This is because degenerative changes or annular dilation of the common AV valve, or both, may progress as the patient ages. [10]

Also Reported mortalities resulting from early primary repair without intervening palliative procedures are generally lower than those reported after two staged repair. [11]

In this context, tight banding can cause severe pulmonary stenosis with myocardial hypertrophy, and loose banding can result in irreversible PVOD. Yamaki and colleagues have reported a patient with VSD who died of PVOD after PAB and subsequent total correction. Therefore, patients who have received PAB should be followed strictly and carefully. The later decision to perform a complete repair is guided by repeat catheterization after PAB and/or lung biopsy. [12]

 By contrast, Prifti and colleagues demonstrated that definitive repairs for small infants who weighed less than 5 kg resulted in an increased risk for late reoperations for residual left AV valve regurgitation. [13]

In all our patients, the choice of the technique used in the repair was based on the surgeon's preference. As a whole in both groups, the Double-patch

technique was the most commonly used technique (26 cases, 65%), then the Modified single-patch technique comes next (8 cases, 20%) and finally comes the Single-patch technique (6cases, 15%).

There has been a recent tendency of many centers to switch from the classic one-patch to the two-patch or the modified one patch techniques. Each technique has its advantage and disadvantages. Surgeon preference and experience with a specific technique will remain the main predictor of choosing the technique. All these techniques seem to be equally efficacious and the ability of the surgeon to adapt to the highly variable pathologic abnormalities of the CAVSD is probably more important than just the technique itself.

The Single-patch technique was the initial technique used for the repair of CAVC defects but was abandoned by many centers due to the difficulty in calculating the height where to attach the divided leaflets to the single patch. Too high or too low level of the left AV valve leaflets create a restricted leaflet motion resulting in a small area of coaptation and subsequent incompetence.[11]

The Two-patch technique has been the technique of choice until recently for most centers .However, the height of the VSD patch should be precisely determined to avoid placing the new left AV valve too high restricting the anterior leaflet in systole. The advantage of the two patches over the classic one patch might be that it avoids division of the valve leaflets, limiting the secondary sequestration of tissue for valve division and reconstruction. Also there is no danger of detachment of the mitral component sutured to the pericardial patch when not dividing the leaflets.[14] [15]

The Modified single-patch technique (the so-called Australian technique) is now becoming more and more popular as an attempt to preserve spatial relationship during repair of common atrioventricular valve by simply attaching the common leaflets to the crest of the septum. The main advantage of not using a VSD patch is that by lowering the level of the left AV valve implantation at the crest of the septum, the area of coaptation is increased resulting in better competence. Another advantage is that it simplifies the procedure reducing both ischemic and total pump times. Wilcox and colleagues reintroduced this method of repair for patients with small VSDs. Nicholson and colleagues advocated that the VSD patch could be avoided in most cases of complete AV canal with moderate and large VSD.[16] [17]

Now progression is going from direct suture of the VSD component, to avoiding the Atrial patch as well, aiming at reducing operating time more and getting normal sized atria that were dilated preoperatively which may help in preventing the occurrence of postoperative arrhythmias, the so-called No-patch technique. One concern with this technique is the risk of applying too much tension on the tissues and subsequent tearing due to the fact of not using any patch .[18]

Conclusion

In conclusion, we have found no difference in operative mortality between patients undergoing surgical repair of CAVSD at 6 months of age or younger. It is true that a difference may not have been surfaced owing to the low statistical power of this study. However, we believe that with modern techniques, the current risk factors for CAVSD repair in patients

younger than 6 months and in those older than 6 months are equal. These factors include the severity of preoperative common AV valve regurgitation, the presence of associated cardiac anomalies, and the degree of functional disability.

Therefore we consider early repair of CAVSD is safe and effective.

As an intermediate strategy that might suit more our circumstances in postoperative care, we propose a policy for the management of CAVCD that is: repair should be done at any age when symptoms of congestive heart failure cannot be managed by medical therapy, while elective repairs for CAVSD are best performed at age 4 to 6 months. In selected cases, as in too low-birth-weight patients, patients with poor clinical condition or with co-morbidities, or in patients in whom complete repair is not feasible due to systemic or higher pulmonary artery pressure initial palliation with pulmonary artery banding may be tried.

References

1-. Najm HK, Coles JG, Endo M, et al. Complete atrioventricular septal defects: results of repair, risk factors, and freedom from reoperation. Circulation 1997;96:II-311–5.

2-Tweddell JS, Litwin SB, Berger S, et al. Twenty-year experience with repair of complete atrioventricular septal defects. Ann *Thorac Surg* 1996; 62:419 –24.

3-Mee RBB, Drummond-Webb JJ. Cardiopatias Congênitas. In: Townsend CM. ed Sabiston Tratado de Cirurgia As Bases Biológicas da Prática Cirúrgica Moderna 2002; Guanabara Koogan: 1370–1371.

4- Castaneda AR, Mayer JE Jr, Jonas RA. Repair of complete atrioventricular canal in infancy. World J Surg 1985;9:590 –7.

5- Hanley FL, Fenton KN, Jonas RA, et al. Surgical repair of complete atrioventricular canal defects in infancy. Twentyyear trends. J Thorac Cardiovasc Surg 1993;106:387–94; discussion 94–7.

6- Manning PB, Mayer JE Jr, Sanders SP et al. Unique features and prognosis of primum ASD presenting in the first year of life. *Circulation* 1994; 90:II30–II35

7- Ramesh R. Singh, Patrick S. Warren, T. Brett Reece, Peter Ellman, Benjamin B. Peeler, and Irving L. Kron. Early Repair of Complete Atrioventricular Septal Defect is Safe and Effective. *Ann Thorac Surg* 2006; 82:1598-1602.

8- Stellin G.,Vidaa V.L., Milanesib O., Rizzolia G., Rubinoa M., Padalinoa M.A.,Bonatoc R., Casarottoa D. Surgical treatment of complete A-V canal defects in children before 3 months of age. *Eur J Cardiothorac Surg* 2003; 23:187-193.

9- Mariko Kobayashi, Yukihiro Takahashi and Makoto AndoIdeal timing of surgical repair of isolated complete atrioventricular septal defect *Interact CardioVasc Thorac Surg* 2007;6:24-26;

10- Wetter J, Sinzobahamvya N, Blaschczok C, et al. Closure of the zone of apposition at correction of complete atrioventricular septal defect improves outcome. *Eur J Cardiothorac Surg* 2000; 17:146 –53.

11- Reddy VM, McElhinney DB, Brook MM, Parry AJ, Hanley FL. Atrioventricular valve function after single-patch repair of complete atrioventricular septal defect in infancy: how early should repair be attempted? *J Thorac Cardiovasc Surg* 1998; 115:1032– 40.

12- Yamaki S, Yasui H, Kado H, et al**.** Pulmonary vascular disease and operative indications in complete atrioventricular canal defect in early infancy. *J Thorac Cardiovasc Surg* 1993; 106:398–405.

13- Prifti E, Bonacchi M, Bernabei M, et al. Repair of complete atrioventricular septal defects in patients weighing less than 5 kg. *Ann Thorac Surg* 2004; 77:1717–26.

14- Weintraub RG, Brawn WJ, Venables AW, Mee RB. Two-patch repair of complete atrioventricular septal defect in the first year of life. Results and sequential assessment of atrioventricular valve function.   *J Thorac Cardiovasc Surg* **1990**; 99:320–326.

15- Bogers AJ, Akkersdijk GP, de Jong PL et al. Results of primary two-patch repair of complete atrioventricular septal defect. *Eur J Cardiothorac Surg* 2000; 18:473–479.

16- Wilcox BR, Jones DR, Frantz EG, Brink LW, Henry GW, Mill MR, Anderson RH. Anatomically sound, simplified approach to repair of complete atrioventricular septal defect. *Ann Thorac Surg* 1997; 64:487–493.

17- Nicholson IA, Nunn GR, Sholler GF, Hawker RE, Cooper SG, Lau KC Cohn SL. Simplified single-patch technique for the repair of atrioventricular septal defect. *J Thorac Cardiovasc Surg* 1999; 118:642–646.

18- Preˆtre R, Dave H, Kadner A, Bettex D, Turina MI. Direct closure of the septum primum in atrioventricular canal defects. *J Thorac Cardiovasc Surg* 2004; 127:1678–1681.